A primer for the student joining the congenital cardiac surgery service tomorrow: Primer 3 of 7

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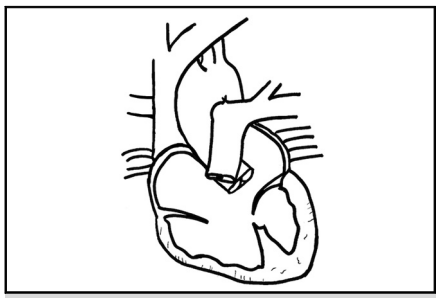

Drawing the anatomy of cardiac defects can help in understanding of the defect and repair.

CENTRAL MESSAGE

This primer prepares students to be acting members of the congenital cardiac surgery service, separated into 3 phases of care: preoperative, perioperative, and postoperative.

Congenital cardiac surgery corrects structural abnormalities of the heart and great vessels.[1] Often, these defects coexist with genetic syndromes and other congenital malformations; therefore, patient management relies heavily on an interdisciplinary approach.[1,2] Congenital cardiac malformations can also be isolated acquired defects.[1] Congenital cardiac surgery services often operate on a wide range of patients, from neonates to adults.[1] It is common for patients to have multiple procedures, which further complicates their surgeries and perioperative management.[3]

This primer covers pediatric congenital heart disease; however, the field continues to evolve, and patients are living into late adulthood. Many patients require reoperations as adults.[3] This primer discusses the nuts and bolts of being on the congenital cardiac surgery service, separated into 3 phases of care: preoperative (clinic and consults), perioperative, and postoperative (patient management). For overall preparation of a cardiac surgery rotation, please refer to the first primer within this series, "A Primer for the Student Joining the Adult Cardiac Surgery Service Tomorrow."

THE EMBRYOLOGICAL BASIS FOR DEFECT

The embryological basis for defect is shown in Table 1.

Order of Embryological Heart Development

1. Formation of the 3 germ layers (gastrulation): ectoderm, mesoderm, and neuroderm.
2. Establishment of the first and second heart fields: primitive streak migration results in a "cardiac crescent" and the establishment of poles.
3. Formation of the heart tube: folding of the germ layers in the craniocaudal and lateral axes brings the endocardial tubes and the lateral plate mesoderm together toward the midline, which fuses into a tube-like structure.
4. Cardiac looping, convergence, and wedging: the 4 chambers of the heart are formed, and spiraling of aortopulmonary trunk takes place (Figure E1).
5. Formation of septa: separation of atrium and creation of the atrioventricular (AV) canals.
6. Development of the outflow tracts.
7. Formation of cardiac valves.
8. Formation of vasculature: coronary arteries, aortic arches, and sinus venosus.
9. Formation of the conduction system.[5-7]

FETAL CIRCULATION

Oxygenated blood is carried from the placenta through the umbilical vein and is partially distributed to the fetal

TABLE 1. Embryological basis for defect[4,5]

Condition	Embryological basis for defect
Persistent truncus arteriosus	Failure of septum formation that divides the truncus arteriosus into aorta and pulmonary artery
Double-outlet right ventricle[1,6]	Incorrect alignment of the aorticopulmonary septum in the truncus arteriosus results in aberrant septation, wherein both great vessels arise from the right ventricle.
Total anomalous pulmonary venous connection (TAPV)	Two theories: (1) The malrotation of the infundibulum prevents clockwise rotation of the aorta toward the left ventricle, which results in displacement of the aorta overriding the right ventricle. (2) Linear rotation instead of spiral rotation of the aortopulmonary septum.
Tetralogy of Fallot (TOF)	Caused by 2 embryological defects: (1) An anterocephalad deviation of the primitive ventricular outflow tract. (2) Abnormal morphology of hypertrophied trabecular muscle that encircles the subpulmonic outflow tract induces right ventricular outflow tract obstruction.
Ebstein anomaly	Failure of leaflet delamination and the continued adherence to the ventricular myocardium causes isolated apical displacement of the tricuspid valve leaflet hinge point with the valve orifice remaining in the normal position. The space between the right ventricle and tricuspid valve hinge point becomes atrialized, leading to varying degrees of tricuspid valve insufficiency.
Hypoplastic left heart syndrome	Failure of delamination or atresia of the mitral valve reduces ventricular filling. The decreased myocardial strain that normally serves as a myocyte growth stimulus results in hypoplasia of the left ventricle.

hepatic circulation, with a majority of blood entering the systemic circulation through the inferior vena cava (IVC), bypassing the liver via the ductus venosus. The blood then travels from the IVC to the right atria, where the direction of flow allows it to cross the foramen ovale into the left atrium. The blood then flows into the left ventricle and into the aorta, which is pumped to the brain (greatest oxygen concentration) or mixed with partially oxygenated blood from the ductus arteriosus, which gets pumped to the rest of the body.[6,8]

Deoxygenated blood from the body comes in through the superior vena cava (SVC) and into the right atria. The downward flow mixes with the oxygenated blood from the IVC and sends partially oxygenated blood into the right ventricle. The partially oxygenated blood enters the right ventricle and is pumped into the pulmonary artery. The blood bypasses the lungs through the ductus arteriosus into the aorta, which mixes with the oxygenated blood to the rest of the body. Deoxygenated blood returns to the placenta through 2 umbilical arteries. A diagram of fetal circulation is shown in Figure 1. Classification of congenital heart defects are listed in Table 2, and common genetic syndromes associated with congenital heart disease are detailed in Table 3.[6,8]

PREOPERATIVE

Diagnosis and Screening: Indications of Congenital Heart Disease

O_2 saturation: <90% in right foot/hand or >3% difference between right foot and hand.

Blood gas analysis + lactate: acidosis and elevated lactate.

Blood pressure: difference >20 mm Hg between arms and legs.[9,10]

Transthoracic Echocardiogram

Doppler echo. *Pulsed wave.*
- Quantifies intracardiac hemodynamics
- Ventricular function

Continuous wave.
- Predicts ventricular pressure
- Mean and peak transvalvular gradients

* Right ventricular pressures/pulmonary artery systolic pressures are estimated using the tricuspid regurgitation velocity, (normal range for peak TR velocity is <2.8 m/s)

Color doppler.
- Direction of blood flow
- Velocity and turbulence
- Identification of shunts

Terminology

Sinus: position of the organs in the body.

- Sinus solitus: normal
- Sinus inversus: mirror image
- Sinus ambiguous: other

Cardiac position: the center of mass of the heart relative to the midline.

- Levoposition: toward the left
- Mesoposition: central
- Dextroposition: toward the right

Apex orientation: the orientation of the base-to-apex axis of the heart.

- Levocardia: toward the left
- Mesocardia: midline
- Dextrocardia: toward the right

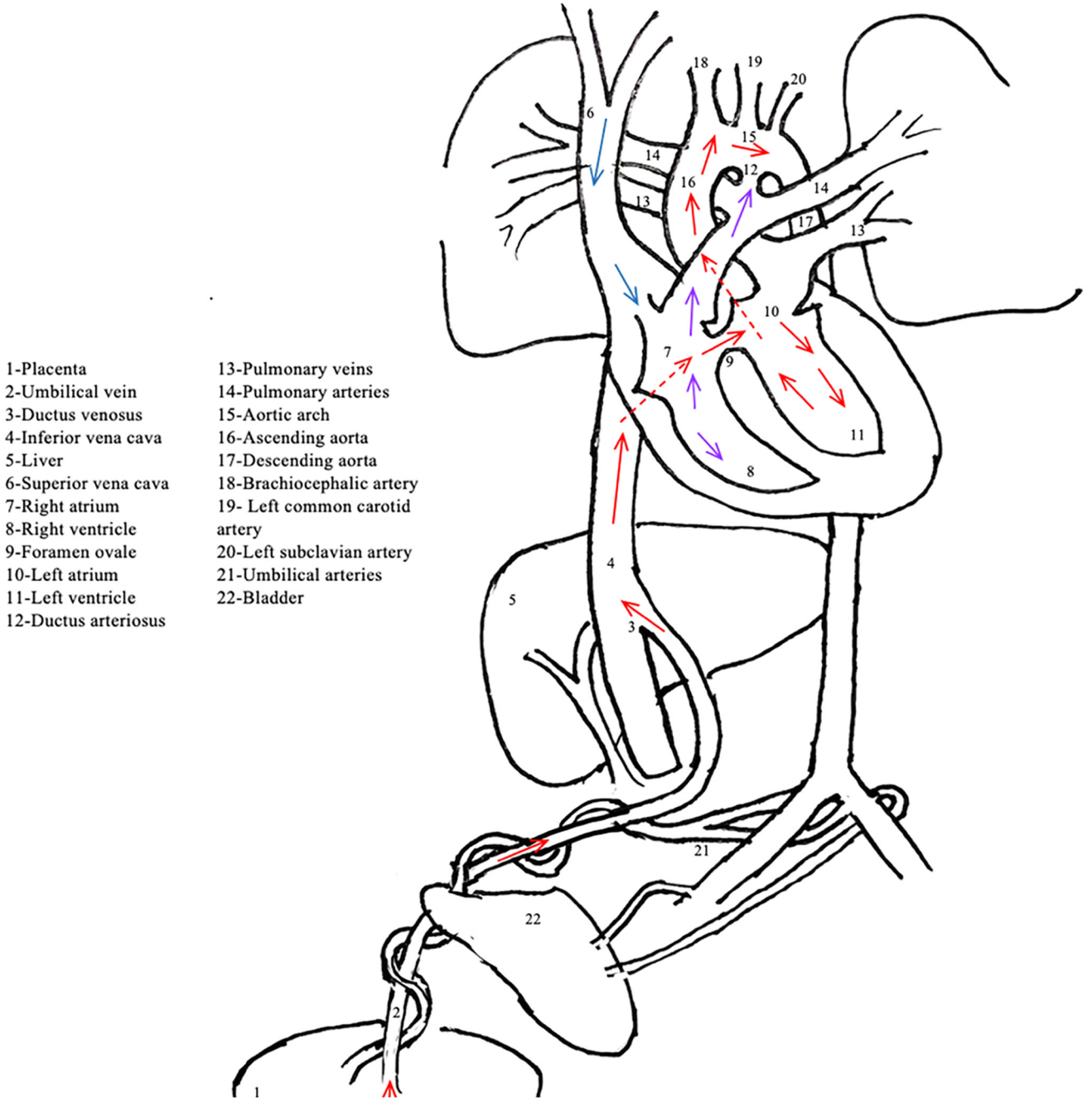

FIGURE 1. Diagram of fetal circulation.

Interpreting Echo Reports

Sequential approach to interpreting echo reports and asssessing cardiac anatomy.

1. Location and position of the heart: sinus, cardiac position, apex orientation.
2. Venous return, atria, and interatrial septum.
3. AV connections and AV valves.

   Asses chamber size (concentric/eccentric hypertrophy).
   Presence of chamber connections.

   Examples:
   Biventricular: one atria is connected to one ventricle.

   - Concordant: normal heart anatomy.
   - Discordant: (corrected transposition).
   - Univentricular: hypoplastic left heart syndrome.

4. Ventricles and the interventricular septum.
5. Outflow tracts and ventriculoarterial connections.
6. Great vessels.

TABLE 2. Classification of congenital heart defects

Cyanotic	
Tetralogy of Fallot	Right ventricular hypertrophy, VSD, overriding aorta, right ventricular outflow obstruction
Transposition of the great vessels	Anatomic reversal of aorta and pulmonary artery
Tricuspid valve atresia	Absent or rudimentary tricuspid valve
Ebstein anomaly	Malformation of tricuspid valve and atrialization of right ventricle
Total anomalous venous return	All pulmonary veins drain into systemic circulation
Persistent truncus arteriosus	Single trunk that receives output from both ventricles
Hypoplastic left heart syndrome	Spectrum of disease with severe hypoplasia of the left ventricle
Acyanotic	
Atrial septal defect	Hole connecting right and left atria
VSD	Hole connecting right and left ventricle
Patent foramen ovale	Failure of atrial septum primum to fuse with septum secundum
Patent ductus arteriosus	Failed closure of the ductus arteriosus
Coarctation of the aorta	Narrowing of the aortic isthmus
Endocardial cushion defect	Defect of the atrioventricular vales

See "Surgical Case Descriptions" section for a more in-depth description of defects. *VSD*, Ventricular septal defect.[1,4,5]

Measurements. Z score: standard deviation above or below an age-specific group mean.

This is useful in serial assessments of disease as a child grows. Normal is within 2 standard deviations.

Ventricular function: left. Ejection fraction.

Fractional shortening.

Left ventricular systolic performance.

Ventricular function: right. Tricuspid annular plane systolic excursion TAPSE: distance tricuspid annulus moves during systole.

Fractional area change: "ejection fraction of the right ventricle."

Example of an echo report. Impression:

1. Large interatrial communication in the canal portion of the atrial septum.
2. Single AV junction.
3. Unrestrictive interventricular communication in the canal portion of the ventricular septum.[11,12]

A complete atrioventricular septal defect (AVSD) is illustrated in Figure 2.

Medical Management as a Bridge to Surgery

The goal of medical management is to reestablish fetal blood circulation with an oxygen saturation goal of 75% to 85%. The approach varies based on the defect.[2,13]

Obstruction and circulation are shown in Table 4.

Important Parameters in Postoperative Management and Transfer to the Pediatric Intensive Care Unit

Respiration.

- What is the respiratory rate and are chest movements symmetrical? Check oxygen saturation and adjust ventilator settings.

Circulation.

- Compare peripheral pulses. If there is an arterial line, check the central venous pressure, cardiac index, systolic blood pressure, pulmonary capillary wedge pressure.

Neurologic status.

- What is their sedation? Check the size of the pupils. Are they spontaneously moving?

Bleeding.

- What is the chest tube output? Are they to suction or water seal?

Temperature.

- Hyperthermia and hypothermia can be signs of sepsis.

Renal function.

- Are they producing urine? Is a Foley in place? Does the color of the urine indicate acute kidney injury? What is the rate of fluid infusion? How are their electrolytes?

Lines/drains/cables.

- Note pacemaker settings. How many lines are there? Are there peripheral lines or central lines? Which side are the lines on?

Important parameters in postoperative management and transfer to pediatric intensive care unit are listed in Table 5. Common postoperative complications and management are shown in Table 6.[2,13]

Surgeon Sign Out

- Diagnosis of the patient and corrective procedure.

TABLE 3. Common genetic syndromes associated with congenital heart disease[4,5]

Congenital heart defect	Associated syndromes
Tetralogy of Fallot	DiGeorge syndrome Trisomy 21
Transposition of the great arteries	Maternal diabetes
Hypoplastic left heart syndrome	Trisomy 13 Trisomy 18 Turner syndrome Jacobsen syndrome
Total anomalous venous return	Heterotaxy syndromes
Persistent truncus arteriosus	DiGeorge syndrome
Tricuspid valve atresia	Trisomy 21

- Surgical techniques and any operative complications.
- Drains and chest tube placement: suction/water seal.
- Intraoperative echo findings.
- Presence of intraoperative bleeding or arrhythmias and management.
- Bypass time.
- Crossclamp time.
- Hypothermia use.

POSTOPERATIVE MANAGEMENT

Fluid Intake

The first 1 to 2 days is resuscitative due to losses in the operating room, vasoplegia from cardiopulmonary bypass (CPB), and capillary leak from CPB. Recommend bolus resuscitation with judicious product use as indicated by coagulopathy/anemia. After day 2 to 3, diuresis can be considered if patient is stable.[1-3,9,13]

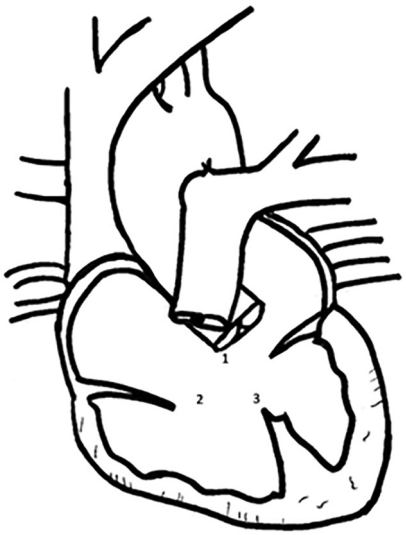

FIGURE 2. Complete atrioventricular septal defect.

Cardiac Dysfunction

Low cardiac output syndrome is a common postoperative complication in patients who underwent procedures requiring CPB. The first step is to evaluate for residual anatomic lesions by echo or cardiac catheterization. Medical management involves milrinone, catecholamines, and pressors. If medical management is not sufficient, mechanical circulatory support is initiated with extracorporeal membrane oxygenation (ECMO) being the preferred method. In extreme cases, an expediated workup for pediatric heart transplant is possible but is complicated by time constraints and anatomical and immunological factors.[1,2,9,13]

Infection prophylaxis

Antibiotics are continued for 2 to 3 days. Patients on ECMO or with open sternotomies require antibiotics until 24 hours following decannulation and/or closure of sternotomy.[1-3,9,13]

Pain Management

A continuous infusion of an opiate and benzodiazepine combination is used in the immediate postoperative period. Pain is then managed with a multimodal regimen, minimizing opiate use to only what is necessary for 2 to 3 days following surgery.[1-3,9,13]

Kidney Function

CBP can lead to renal impairment, and acute kidney injury is very common after cardiac surgery. Urine excretion of at least 2 mL/kg/h after cardiac surgery is the minimum to ensure adequate fluid balance. In addition, loop diuretics are administered after the resuscitative phase.[1-3,9,13]

SURGICAL CASE DESCRIPTIONS (TYPES, INDICATIONS, AND STEPS)

Patent Ductus Arteriosus (PDA)

The ductus arteriosus is a normal part of fetal circulation that arises from the distal portion of the sixth and eighth embryonic aortic arch. In a PDA, the communication remains patent between the systemic and pulmonary vasculature. The most common location of a PDA is between the isthmus of the distal aortic arch and the origin of the left pulmonary artery.[1,3,13,14]

Clinical Features

Small PDA

- Asymptomatic.

Large PDA

- Failure to thrive.
- Heaving laterally displaced apical pulse (left ventricular volume overload).

TABLE 4. Obstruction and circulation

Left heart obstruction	Relies on patent duct for systemic circulation	Hypoplastic left heart syndrome, critical aortic stenosis/ coarctation
Right heart obstruction	Relies on patent duct for pulmonary circulation	Critical pulmonary valve stenosis/atresia, severe tetralogy of Fallot, severe Ebstein anomaly
Parallel circulation	Relies on duct for pulmonary and systemic circulation	Transposition of the great arteries
Approach to ductal-dependent systemic circulation: 1. Provide patency of ductus arteriosus with continuous prostaglandin E1 infusion 2. Lower systemic vascular resistance - Reduce afterload with sodium nitroprusside infusion - Initiate milrinone and/or dobutamine 3. Increase pulmonary vascular resistance - Avoid intubation; aim for mild metabolic acidosis and mild hypoventilation		Approach to ductal-dependent pulmonic circulation 1. Provide patency of ductus arteriosus with continuous prostaglandin E1 infusion 2. Lower pulmonary vascular resistance - Aim for mild metabolic alkalosis, hyperventilation, and increase inspired oxygen fraction 3. Increase systemic vascular resistance - Initiate norepinephrine or epinephrine - Increase volume status

- Wide pulse pressure with bounding pulses.
- Loud continuous machinery murmur heard best at the left infraclavicular region.
- Symptoms of pulmonary overcirculation, feeding intolerance, dyspnea.

Indication for Operation

Initial management includes COX inhibitors, with indomethacin being the preferred pharmacological treatment to close the PDA. Operative management is indicated in patients who are unresponsive to medical management and who have hemodynamically significant PDAs. Operative interventions include a percutaneous or surgical approach.[1,3,13,14]

Surgical ligation is performed in patients who have the following:

- large PDA;
- refractory or worsening ventricular dependence;
- poor tissue perfusion;
- failed trial of COX inhibitors; or
- contraindication to percutaneous closure (such as prematurity).

Most Common Operation: Double Ligation and Clip Technique/Ligation With Division

Open procedure is the standard technique and can be performed in neonates >350 g. In infants born premature with low birth weights, closure is done with suture ligation or hemoclip without division.[1]

1. Create a left posterolateral thoracotomy incision (*avoid the serratus anterior and latissimus dorsi*). Enter into pleural space through the fourth intercostal space.
2. Identify recurrent laryngeal nerve to avoid injury.
3. Place hemoclip at the aortic end of the PDA.
4. Place the second suture around PDA through the ductus adventitia closest to the pulmonary artery (*This is the last step of ligation and clip technique for premature infants*).
5. Ligation with division: After identification and mobilization of the PDA, place 1 to 2 hemoclips across the ductal origin (*avoid vagus and recurrent nerves*).[1,3,14]

Notes

Other options in infants born premature with low birth weights:

- Single hemoclip.
- Single ligation.
- Combination of ligation and single hemoclip.

Complications. Pneumothorax, chylothorax, bleeding, aortic obstruction, pulmonary artery obstruction, residual flow, vocal cord dysfunction, wound infection, blood pressure fluctuations, respiratory compromise, infection, intraventricular hemorrhage, bronchopulmonary dysplasia, and death.[1,3,13,14]

Coarctation of the Aorta (Isolated) in the Neonate

A form of left ventricular outflow obstruction caused by an abnormal narrowing of the aorta. It is most commonly found at the juxtaductal aortic isthmus just distal to the left subclavian artery.[1,3]

Clinical Features

- Differential cyanosis of the lower extremities.
- Brachial–femoral delay with weak femoral pulses.
- Cold feet and lower-extremity exertional claudication.
- Continuous blowing murmur best heard below the left clavicle or between shoulder blades.
- Systolic ejection murmur over left posterior hemithorax.

TABLE 5. Important parameters in postoperative management and transfer to PICU

Ventilator settings		
Respiratory rates, breaths/min Neonates: 40 Infants: 25-30 Toddlers: 25 Elementary school–aged children: 20 Adolescents: 15	Tidal volume: 6-8 mL/kg PEEP: 5-10 mm Hg FIO_2: pathology dependent	Variants: Univentricular heart: target oxygen saturation 80% Fontan heart: PEEP <5 mm Hg Pulmonary HTN: Mild hyperventilation FIO_2 level for O_2 goal of 100%, PaO_2 >150 mm Hg

Hemodynamics	Normal range
Cardiac output, L/min	
Neonate/infant	0.8-1.3
Child	1.3-3.0
Cardiac index, L/min/m^2	
Neonate/infant	4.0-5.0
Child	3.0-4.5
Heart rate, beats/min	
Neonate/infant	100-180
Pediatric	70-110
Stroke volume, mL/beat	
Neonate/infant	5-13
Pediatric	13-50
Preload RAP/CVP, mm Hg	
Neonate/infant	0-8
Pediatric	2-6
Right atrium, infant	1 mm Hg
Right ventricle	26/2 mm Hg
Pulmonary artery, infant	26/12 mm Hg
Left atrium, infant	3 mm Hg
Left ventricle, infant	60/3 mm Hg
Aorta, infant	70/50 mm Hg

PEEP, Positive end-expiratory pressure; *FIO_2*, inspired oxygen fraction; *HTN*, hypertension; *PaO_2*, arterial oxygen tension; *RAP*, right atrial pressure; *CVP*, central venous pressure; *PICU*, pediatric intensive care unit.[2,13]

Indications for Operation

Corrective intervention by surgery or transcatheter intervention should be performed in all patients with critical coarctation. This includes transaortic gradient >20 mm Hg (*gradients might not be present in patients on prostaglandin [PGE]), significant collateral flow, and or/symptomatic hypertension or heart failure. Preoperative management involves securing patency of PDA to improve distal perfusion with PGE1 infusion. Surgical intervention in the preferred operative management, but percutaneous dilation through balloon angioplasty or stent placement can also be performed.[1,3]

Most Common Operation: Extended End-to-End Anastomosis Without CPB

1. Begin with a posterolateral thoracotomy with entry into the thoracic cavity through the fourth intercostal space.
2. Dissect and reflect pleura overlying transverse and descending aorta (*Identification of phrenic and recurrent laryngeal nerves at this step is crucial to avoid injury*).
3. Dissect the transverse aorta, descending aorta, and ductus arteriosus. The goal is to reduce any tension to prepare for later anastomosis.
4. Place a vascular clamp across the transverse aorta, distal to the brachiocephalic artery. The clamp should also clamp off the left carotid and subclavian arteries.
5. A second clamp is placed across the descending aorta past collateral arteries.
6. Ligate and remove collateral arteries.
7. Place a suture around the PDA for hemodynamic control and mobility. Excise the narrowed section of the aorta.
8. Reconnect the free ends of the aorta using a running monofilament suture. Begin at the apex of the transverse arch and suture posteriorly to the descending aorta.

TABLE 6. Common postoperative complications and management

Low cardiac output syndrome	Replace volume, inotropes, afterload reduction (ie, sodium nitroprusside)
Pulmonary hypertensive crisis	Administer O_2, alkalosis, inhaled NO, hyperventilation
Tachyarrhythmias: beta-blockade, amiodarone, cardioversion if unstable	
Stable re-entrant tachycardia	Vagal maneuvers, adenosine, or beta-blockers
Unstable re-entrant tachycardia	Adenosine, synchronized cardioversion
Ectopic atrial tachycardia	Beta-blockers (eg, esmolol)
Junctional ectopic tachycardia	Amiodarone
Bradyarrhythmia: pacing, atropine in the emergent setting	
Isolated sick sinus syndrome	Temporary pacing wires with atrial pacing
AV block	Temporary pacing wires with AV sequential pacing

NO, Nitric oxide; *AV*, atrioventricular.[2,13]

Notes

Both balloon angioplasty and stent placement have increased risks of aortic aneurysms, reoccurrence, and the need for additional surgical corrections later in life. Previous balloon angioplasty is also associated with a greater risk of paraplegia due to surgical complications in patients who require failed balloon angioplasty revision surgery. It is therefore reserved for recurrent coarctation after surgical intervention, which occurs in up to 20% of patients.[1,3]

Complications. Paraplegia, chyle leak, and recurrent nerve injury.

Atrial Septal Defect (ASD)

An ASD is a hole in the septum connecting the left and right atria. It occurs due to impaired growth or excessive resorption of the atrial septum in utero. Most commonly, they are minor left-to-right shunts with low-volume flow across the defect. If left untreated, more severe defects can lead to arrhythmias, pulmonary hypertension, a right-to-left blood flow reversal shunt (Eisenmenger syndrome), and right ventricular failure/dysfunction. The most common type of ASD is an isolated ostium secundum (70%). These defects are centrally located. An ostium primum ASD is located in the lower portion of the septum and often co-occurs with other cardiac defects, such as mitral and tricuspid valve defects. Other less common ASD locations include a sinus venosus defect between the SVC and right-sided pulmonary veins and an "unroofed" coronary sinus defect.[1,3,15,16]

Clinical Features

Small (<5 mm)—asymptomatic

Medium to large (8-10 mm)

- Systolic ejection fraction best heard at the second intercostal space at the sternal border.
- Systolic ejection fraction best heard at the second intercostal space at the sternal border.
- Wide fixed split S2.
- Soft mid-diastolic murmur over the lower left sternal border (increased flow across the tricuspid valve).

Indications for Operation

Closure is recommended between 2 years and 4 years of age in patients with significant left-to-right shunt flow, symptomatic right-sided volume overload, pulmonary 1.5:1, or heart failure. Closure involves either a transcatheter or surgical approach. Surgical operations are indicated in complicated ASDs, defects >38 mm, or defects that have deficient muscular rims. Small ASDs or patent foramen ovales (PFOs) can be closed by direct suture; larger defects require a patch.[1,3,15,16]

Most Common Operation: Ostium Secundum ASD Closure Using Autologous Pericardial Patch

1. Begin with standard sternotomy, limiting skin incision size as much as possible.
2. Dissect and reflect the thymus for aortic cannulation and initiate cardiopulmonary bypass.
3. Open pericardium left of the midline.
4. Perform a right atriotomy. Place sutures around the edges of the atriotomy to facilitate traction and to visualize ASD.
5. Locate the AV node at the superior apex of the triangle of Koch. The triangle can be identified by the position of the coronary sinus relative to the tricuspid valve and the tendon of Todaro.
6. Confirm the position of pulmonary veins and IVC to prevent injury.
7. Place sutures at the superior and inferior border of ASD then thread the sutures through a measured and cut pericardial patch to align correctly on ASD.

8. Suture in place using multiple interrupted pledgeted mattress sutures or with a running monofilament suture.
9. Leave a small gap in the patch to perform deairing maneuvers. Deair by ventilating the patient (this increases pulmonary return and volume in the left atria to remove any remaining trapped air). Once there is confirmation that there is no remaining air, the remainder of the patch can be closed.
10. Begin rewarming while closing left atriotomy. Wean from CPB once rewarming is complete.[1,3,15,16]

Notes

Complications. Heart block, insufficient closure, iatrogenic right-to-left shunt.[1-3]

Ventricular Septal Defect (VSD)

VSDs are the most common congenital heart defect, with an incidence in approximately 4 in 1000 live births. It can occur as an isolated defect or concurrently with other congenital heart abnormalities. In some cyanotic heart defects, its presence is necessary for survival. The most common location of VSDs occur in the ventricular septum's membranous portion, just beneath the aortic valve and behind the septal leaflet of the tricuspid valve. Defects can also occur in the muscular portion of the septum and be centrally or apically located along the right ventricular free wall-septal junction. Other less common defects include malalignment defects, subpulmonic defects, and AV canal inlet defects.[1,3]

Clinical Features

Small (<5 mm). Murmur can be heard at approximately 4 to 10 days of life, when the pulmonary vascular resistance decreases. This lowers right ventricular pressures and allows for left-to-right shunting.[1-3]

- Asymptomatic.
- Harsh holosystolic murmur best heard at the left lower sternal border (LLSB).
- Murmur increases in intensity with increased afterload (eg, hand grip).

Medium to large (6-10 mm). If left untreated, symptoms of heart failure typically present by 3 to 4 weeks of life.

- Harsh holosystolic murmur best heard at the LLSB (no murmur in large VSDs).
- Failure to thrive.
- Hepatomegaly.
- Pulmonary rales, grunting, and retractions.
- Tachycardia.
- Mid-diastolic murmur over cardiac apex.
- Systolic thrill.
- Loud pulmonic component of S2 from pulmonary hypertension.

Indications

Small defects typically close on their own or decrease in size over time; they typically do not require intervention. The goal of medical management in symptomatic patients is to reduce symptoms of heart failure before surgical correction. Closure is indicated in children <1 year with symptoms of pulmonary hypertension (Qp:Qs > 1.5), heart failure, right ventricular outflow obstruction, and/or left ventricular dilation.[1,3]

Most Common Operation VSD Closure With Patch

1. Initiate CBP.
2. Expose through atriotomy.
3. Line the VSD with superficial 5-0 or 6-0 interrupted or continuous sutures.
4. Run sutures through a polytetrafluoroethylene (ePTFE) patch and lower into VSD to complete repair.
5. Perform deairing maneuvers and close atriotomy.
6. Wean off bypass.

Notes

The most common approach is through a right atriotomy but could differ based on the location of the defect. An important takeaway is to take small bites during repair to avoid injury to a valve or the conduction system.[1,3]

Complications. AV block.

Damage to the tricuspid valve, pulmonic valve, or aortic valve.

Incomplete closure of the VSD.[1-3]

AVSDs (AV Canal)

A broad range of AVSDs occur due to a partial or complete failed fusion of the endocardial cushions. This results in defects in the AV valves and the atrial and/or ventricular septum. There is a high association between AVSDs and trisomy 21, with 40% to 50% of neonates with AVSDs having concurrent Down syndrome. AVSDs are categorized into complete, partial, transitional, or intermediate forms. Complete AV canal defects (CAVCs) have a single AV valve often associated with significant regurgitation. The left AV valve is most commonly affected in partial AV canal defects.[1,3,10,12]

Clinical Features

Complete. *Associated with heart failure findings.* Hyperactive precordium with inferior and laterally displaced precordial impulse.

Increased pulmonary component of the second heart sound (S2) due to pulmonary artery hypertension.

Systolic ejection murmur heard at the left upper sternal border (increased blood flow across the pulmonary valve).[1,3,10,12]

Partial. *Associated with clinical features similar to ASD. Often, they are asymptomatic but can develop symptoms of heart failure if there is significant mitral regurgitation or pulmonary circulation overload.* Wide and fixed splitting of the S2 during respiration.

Systolic ejection murmur at the left upper sternal border with radiation to the lung fields (increased blood flow across the pulmonary valve).

Increased right ventricular precordial impulse.

Diastolic rumble (increased blood flow across the tricuspid valve).

Holosystolic murmur (related to mitral regurgitation).[1,3,10,12]

Indications

Surgical repair for AVSDs is almost always required for both complete and partial defects. Most children with CAVCs will develop heart failure by 1 year of life if left untreated. Medical management for symptoms of heart failure is indicated before surgery. CAVC repair should occur in early infancy before the age of 2 to 3 months. In partial defects, repair typically occurs between 18 months and 3 years. The choice of operation is often institution- and patient-dependent.[1,3,10,12]

Two-Patch Technique

1. Initiate aortocaval CPB and cardioplegia.
2. Approach through a right atriotomy.
3. Visualize the extent of the interventricular septum by incising the common posterior leaflet.
4. Divide the superior and inferior bridging leaflets of the common AV valve.
5. VSD closure: Place individual pledged sutures into the right side of the interventricular septal crest. (*avoid conduction system). Lower a pericardial patch onto the defect and anchor to the septum with pledgeted sutures.
6. Repair the AV valves first by dividing the superior and inferior bridging leaflets.
7. Approximate the leaflets and attach them to the common patch using an interrupted suture technique.
8. ASD closure: Lower a section of the pericardium to approximate size needed for ASD closure. Suture pericardial patch to the edges of the ASD in the left atrium adjacent to interatrial septum (*careful attention is used to avoid damage to the conduction system which lies near the coronary sinus*).
9. Connect AV valve leaflets to the top of the ventricular septum.
10. Test AV-valve function with a saline bulb syringe.[1,3]

Modified One-Patch Technique (Nunn Repair)

1. Initiate aortocaval CPB and cardioplegia.
2. Approach through a right atriotomy.
3. VSD closure: Place individual pledged sutures into the right side of the interventricular septal crest (*avoid conduction system). Lower a pericardial patch onto the defect and anchor to the septum with pledgeted sutures. This separates the AV valve into left and right components.
4. Repair the AV valves by approximating the leaflets and attaching them to the common patch using an interrupted suture technique.
5. ASD closure: Close ASD using a running suture.
6. Test AV valve function with a saline bulb syringe.
7. Complete the procedure by performing deairing maneuvers. Close atriotomy and wean off of bypass.[1,3]

Notes

Other operations. *One-patch technique.* One patch is used to divide atrial and ventricle communications. The common AV valve is then secured to the patch. This is an uncommon approach due to difficulty attaching leaflets to the patch and the creation of residual VSDs.[1,3]

Two-patch technique. VSD is closed using a patch with the common AV valve sutured on the superior edge. A second patch is used to close the ASD and then it is secured to the common AV valve and ventricular patch.[1,3]

Partial AVSD: patch repair of ostium primum and mitral valvuloplasty. For partial AV canal defects, the ventricular septal defect may be closed primarily or with a patch, depending on its size.[1,3]

Complications. High risk of AV block.

Residual septal defect.

Valve regurgitation.[1-3]

Persistent Truncus Arteriosus (PTA)

The etiology of a PTA is an underdeveloped aorticopulmonary septum. The truncus arteriosus (the common arterial trunk) fails to divide into a separate pulmonary trunk and aorta. This results in blood being pumped through the heart through a single truncal valve into a single truncal artery. The trunk provides blood for the entire systemic, pulmonary, and coronary circulatory systems. Older classifications of PTA subtypes include the Collet Edwards and Van Praagh classifications. The anatomy of the pulmonary vasculature categorizes the former, and the latter is further subdivided by aortic abnormalities. The Society of Thoracic Surgeons modified the classifications into 3 categories:

1. TA with confluent or near confluent pulmonary arteries.
2. TA with an absence of one pulmonary artery.
3. TA with interrupted aortic arch or coarctation.[1,3,17]

Clinical Features

Most neonates present within the first weeks of life with cyanosis, pulmonary congestion, and heart failure.

- Failure to thrive.
- Cyanosis.
- Respiratory distress and pulmonary edema.
- Bounding peripheral pulses.
- Harsh systolic murmur at LLSB.
- Loud S2.

Indications

Surgical correction is always indicated to prevent early mortality. If left untreated, the 1-year survival is approximately 15%. Medical management with diuretics, inotropes, angiotensin-converting enzyme inhibitors, and noninvasive positive pressure ventilation is required before surgery to stabilize cardiopulmonary function.[1-3]

Most Common Operation: Repair of Simple Truncus Arteriosus

In the setting of functional truncal valve:

1. Approach through median sternotomy. Open the pericardium and mobilize the pulmonary arteries with tourniquets for control.
2. Initiate CPB (*Snares are placed on pulmonary arteries to prevent pulmonary overcirculation during cooling).
3. Crossclamp the aorta as distally as possible to allow for sufficient space during the division of the pulmonary arteries from the truncal vessel.
4. Separation and repair of the pulmonary trunk from the common trunk: Excise near the origin of the pulmonary bifurcation on the posterior side of the common trunk (*This step requires careful attention to avoid injury to coronary vessels*). Repair the common trunk (which is now the neoaorta) using a patch (pericardial patch or a pulmonary or aortic homograft can be used). Mobilize the pulmonary bifurcation to the left side for reconstruction.
5. Closure of the VSD: Approach through right ventriculotomy and close defect with a patch and a running suture.
6. Right ventricular outflow tract (RVOT) reconstruction: Create a conduit between the right ventricle and pulmonary arteries using an aortic allograft. Anastomose the pulmonary homograft and the pulmonary bifurcation with a running monofilament suture.
7. If ASD is present, a partial closure is done to allow for right to left shunting in the case of postoperative ventricular dysfunction.
8. Rewarm and wean off bypass (*Snares are placed on the right and left pulmonary arteries to prevent pulmonary overcirculation during warming).[1,3]

Notes

Complications. AV block.

Residual VSD.

Truncal valve regurgitation.

Right ventricular dysfunction/poor right ventricular output if poorly placed right ventricle to pulmonary artery conduit.[1-3]

Tetralogy of Fallot (TOF)

TOF is the simultaneous occurrence of subpulmonic infundibular stenosis (RVOT obstruction), right ventricular hypertrophy, VSD, and an overriding aorta. The severity of cyanosis and physiological consequences are closely associated with the degree of RVOT obstruction. RVOT obstruction forces a right-to-left shunt through the VSD, resulting in a large volume of desaturated blood entering the systemic circulation. Transient hypercyanosis, known as a tet spell, is a transient near-complete occlusion of RVOT and occurs during periods of stress. Squatting or bringing the knees to the chest can alleviate symptoms. Prolonged spells can lead to loss of consciousness and cardiac arrest.[1,3,18]

Clinical Features

- Tet spells: hypercyanotic hypoxic episodes with peak incidence between 2 and 4 months.
- Harsh crescendo decrescendo systolic ejection murmur at the left upper sternal border.
- Single S2.
- Right ventricular heave and palpable thrill.
- Symptoms of congestive heart failure.

Indications

Complete surgical repair is done within the first year of life. Immediate palliative shunt or ductal stent may be required in infants with severe RVOT obstruction who are ductal-dependent. Repair in severe obstruction is done within the 4 weeks of life. Constant PGE1 infusions are necessary until the time of operation. Infants with less-severe RVOT obstruction receive elective surgery between 2 and 4 months when pulmonary vascular resistance has decreased. There are variations in the procedure based on the severity of RVOT, which is determined by a z score. In some cases, the RVOT is not severe enough to warrant enlargement of the pulmonary valve. In addition, if the pulmonary valve annulus is severe enough to need correction, but not too narrow to need total repair, a transannular patch will be used to enlarge the RVOT.[1-3,18]

Most Common Operation: Complete Repair of TOF

1. Approached through a midline sternotomy. Initiate bicaval cannulation CPB.

2. Correct RVOT obstruction: create an incision at the base of the right atrial appendage and retract the anterior tricuspid leaflet to expose the infundibulum.
3. Resect infundibular muscle bundles using an inverted Y incision. The resection should create an outflow tract that is 2 mm larger in circumference than the pulmonary annulus.
4. Enlarge pulmonary valve: Perform careful commissurotomies to enlarge the pulmonary valve orifice without sacrificing the integrity of the valve.
5. VSD closure: Using a transatrial approach, close VSD with a polytetrafluoroethylene (PTFE) patch and interrupted, pledgeted sutures. Use Teflon-felt pledgets to prevent tearing through the tissue of the right interventricular septum (*Avoid injury to the tricuspid valve apparatus, the aortic valve, and the conduction system*).
6. Probe through the RVOT to ensure adequate pulmonary valve annulus diameter. Remove any air from the left heart.
7. Close right atrium.
8. Reconstruct exterior RVOT: Use an autologous pericardial patch and interrupted sutures to complete supravalvular repair.[1,3,18]

Notes

Complications. Conduction block.

Insufficient RV muscle bundle resection.

Residual VSD.

Insufficient patch repair of pulmonary artery, residual stenosis.[1-3]

Transposition of the Great Arteries (TGA)

In TGA, the aorta arises from the right ventricle, and the pulmonary artery arises from the left ventricle. This occurs due to failed spiraling of the aortopulmonary septum. The result is 2 parallel circulatory loops in complete isolation. The right ventricle receives and pumps deoxygenated blood into systemic circulation, bypassing the lungs. The left ventricle receives and pumps blood through the pulmonary circulation, bypassing the body. The condition is only survivable if there is intracardiac mixing of blood through septal defects or extracardiac mixing through a PDA.[1-3]

Clinical Findings

- Tachypnea.
- Single loud S2.
- Diminished femoral pulses.
- Murmur may be present with concurrent VSD.

Indications

Timing and type of operation are largely determined by the nature of the cardiac abnormalities. There are 3 most common presentations of TGAs. The most common type (50%) is TGA with an intact interventricular septum. Initial management involves balloon atrial septostomy for hemodynamic stabilization. An arterial switch operation (ASO) is performed within the first 2 weeks of life. The second type (25%) is TGA with a concurrent VSD. A transcatheter balloon septostomy may or may not be indicated. The operation of choice is an ASO and closure of VSD within the first 2 weeks of life. The third type (25%) is TGA with pulmonary stenosis. Initial management involves systemic-to-pulmonary artery shunt followed by a Rastelli operation later in life.[1-3]

Most Common Operation: ASO With and Without VSD

1. Place patient on CPB and administer cardioplegia.
2. Repair ASD: Approach through atriotomy and close defect using a pericardial patch.
3. Repair VSD (if present): Approach through the right atrium. Repair defect using a pericardial patch and a running suture.
4. Translocate aorta: Retract and transect aorta away from the main pulmonary artery.
5. Transfer of coronary arteries: There are 2 techniques to mobilize and reanastomose the coronary arteries, the open trap door or closed (punch technique). Anastomose the coronary arteries to the pulmonary artery—this will become the new aortic root (*Delineate any aberrant coronary artery anatomy before surgical correction*).
6. Coronary button anastomosis is done using 8-0 or 7-0 running monofilament suture on one side. Individual sutures are then placed at the top of the anastomosis to anchor the running suture.
7. LeCompte maneuver: Translocate the pulmonary artery so that the bifurcation is anterior to and straddling the aorta. (*The maneuver ensures proper alignment while simultaneously reducing tension. It decreases the risk of subsequent pulmonary artery stenosis and reintervention rates*). The neoaortic anastomosis is performed with a running suture technique.
8. Begin rewarming the patient. Perform deairing maneuvers and release crossclamp before the neoaortic anastomosis is complete.
9. Neopulmonary artery reconstruction: Use a pericardial patch to reconstruct and fill in the gaps of the neopulmonary artery where the coronary arteries were removed. Secure with 7-0 monofilament running sutures (*Make sure the pericardial patch is tapered as a straight patch can lead to supravalvular pulmonary stenosis*),
10. Close patient and wean off bypass.[1,3]

Rastelli Procedure

Indicated for TGA associated with a large VSD and left ventricular outflow obstruction. The goal of this procedure is to augment the VSD so that blood flow is directed from the left ventricle to the aorta. The right ventricle is connected to the pulmonary artery through an external conduit. The Rastelli procedure requires a large intrinsic VSD. The VSD can be surgically enlarged, but this carries the risk of complete heart block and reliance on a pacemaker. Other operations to correct TGA are the Nikaidoh or the REV procedure, which do not require a conduit between the pulmonary artery and right ventricle.[1-3]

1. Initiate CPB and cardioplegia.
2. Ventriculotomy is performed away from the course of the left anterior descending coronary (*Avoid damage to the aortic valve, which lies anterior and is subject to injury*).
3. Infundibular resection: perform a scissor dissection keeping the aortic valve in view at all times. A dilator may be placed through the aorta to guide the resection.
4. Resect the subaortic obstructing mass.
5. Baffling of the LVOT through the VSD: baffle the LVOT with a patch and interrupted, pledgeted sutures. This will establish an outflow tract from the left ventricle to the aorta.
6. Optional: transect the aorta and insert a dilator through the aorta into the left ventricle to ensure there is no stenosis created by VSD patch.
7. Rewarm patient.
8. Create a right ventricle conduit to the pulmonary artery using an extracardiac valve conduit.
9. Wean off bypass and close.

Notes

Complications. LV outflow tract obstruction and LV hypertrophy.

Conduction block.

Coronary button problems with subsequent ischemia.

Damage to the valves.[1-3]

Total Anomalous Pulmonary Venous Return (TAPVR)

TAPVR is characterized by the failure of the pulmonary veins to connect to the left atrium. The pulmonary veins, instead, connect to the right atrium or its tributaries. Intracardiac blood mixing is required for survival. The resulting pathology includes venous mixing, pulmonary venous obstruction, and left-to-right shunting. The degree of disease severity depends on the extent of pulmonary venous obstruction. There are 4 anatomic variants:

Type 1 (45%-50%) supracardiac wherein the pulmonary veins drain into the brachiocephalic vein or SVC.

Type 2 (15%-20%) intracardiac, where the pulmonary veins drain into the coronary sinus.

Type 3 (20%-25%) infracardiac with drainage into the IVC or portal vein.

Type 4 (10%), mixed type, which consists of a combination of pulmonary drainage and is typically the most challenging to correct.[1-3,19]

Clinical Features

Infants with severe pulmonary venous obstruction present as critically ill with symptoms of respiratory failure and shock. Unobstructed infants have subtle cyanosis that is usually detected by pulse oximetry.[1-3]

- Cyanosis.
- Respiratory failure.
- Failure to thrive.
- Hepatosplenomegaly from right-sided heart failure.
- Fixed split S2 or loud S2 if an obstruction is present.
- Systolic ejection murmur with diastolic rumble if no obstruction is present.

Indications

Infants with severe obstruction require medical management with PGE1 and respiratory support until they undergo surgical correction. Additional palliative interventions may include ECMO and temporizing cardiac catheterization. Surgery is indicated in all patients regardless of the degree of obstruction and is performed at the time of diagnosis or once stabilized. The type of operation is determined by the anatomical variant. The goal of surgery is to reestablish a direct pathway between the pulmonary veins and the left atrium without obstructing pulmonary venous drainage. **Obstructed TAPVR is the one true emergency of congenital cardiac surgery.[1-3]

Most Common Operation

Repair of type I (supracardiac) total anomalous pulmonary venous connection: posterior approach.

1. Initiate CPB (over a period of 20-25 min) and deep hypothermia (18 °C).
2. During cooling, the PDA is ligated. The cardinal vein, brachiocephalic artery, and confluence of pulmonary veins are dissected (*special care is taken to avoid injury to the phrenic nerve and recurrent nerve during this dissection*).
3. Initiate cardioplegia once the target temperature is reached.
4. Closure of ASD: Approach through left atriotomy. PFOs are often located in the superior part of the left atrium in TAPVR.
5. Align the confluence of pulmonary veins with the left atrium in preparation for anastomosis. Perform a side-to-side anastomosis. Place the first suture at the right-

sided pulmonary vein. Use a running suture technique with retraction and segmental release. Ligate the vertical vein (*Make sure not to confuse the pulmonary vein with the vertical vein during ligation).
6. Close and rewarm patient. Wean off of bypass.[1,3]

Repair of type II (intracardiac) total anomalous pulmonary venous connection. These types rarely have pulmonary obstruction and present with similar hemodynamics to ASD.

1. Initiate CPB and cardioplegia.
2. Approach through a right atriotomy.
3. Identify the collecting vein of the coronary sinus into which the pulmonary veins drain.
4. Enlarge and unroof the coronary sinus up to the mitral valve.
5. Repair ASD with a pericardial patch (*Make sure not to obstruct pulmonary veins).
6. Close and wean the patient off of bypass.[1,3]

Notes

Complications. Pulmonary venous stenosis is the most feared complication. Additional complications may include incomplete PFO closure and accidental division of the pulmonary veins (which can sometimes be mistaken for a vertical vein).[1-3]

Hypoplastic Left Heart Syndrome (HLHS)

HLHS is characterized by severe hypoplasia of the left ventricle. There are also variations in which there is stenosis or atresia of the mitral valve, aortic valve, or aortic arch. In HLHS syndrome, the left ventricle is nonfunctional and the right ventricle is the primary supplier for both pulmonary and systemic circulations. Survival is dependent on the presence of a PDA for systemic perfusion from right atrium to aorta, and an ASD to allow for adequate mixing of oxygenated and deoxygenated blood. If left untreated, 95% of neonates will not survive the first few weeks of life.[1,3]

Clinical Features

- Cyanosis.
- Tachypnea.
- No murmur.
- Cool extremities.
- Hepatosplenomegaly.
- Dysmorphic features indicative of an underlying syndrome.

Indications

All infants born with HLHS require surgical correction for survival. A cardiac transplant is curative, however, there are limited infant donors. The more common approach is staged surgical revision through a series of 3 different procedures in early life. The staged procedures do not restore the patient to normal biventricular circulation. For illustrations of procedures, please refer to the following article: https://www.frontiersin.org/articles/10.3389/fped.2020.627660/full

https://www.chop.edu/treatments/staged-reconstruction-heart-surgery.[1,3,19]

Stage 1 Norwood procedure: **occurs in the first few days of life.** The Norwood procedure reconstructs and enlarges the aorta through an aortopulmonary amalgamation and patch augmentation. It is a type of Damus–Kaye–Stansel anastomosis that merges the aorta and pulmonary artery to create a functional right ventricular outflow tract to the systemic circulation. A shunt is then used to connect the brachiocephalic artery and the right pulmonary artery to establish pulmonary circulation. The final step is an atrial septectomy if intrinsic ASD is not large enough to allow for adequate mixing of blood.[1,3]

There are 2 types of perfusion strategies used in the Norwood operation: deep hypothermia and circulatory arrest and regional perfusion techniques without circulatory arrest. They can also be used interchangeably throughout the procedure.[1,3]

Stage 2 Glenn procedure: **occurs between 3 and 6 months.** Reattaches SVC from the heart to the pulmonary artery.

Stage 3 Fontan procedure: **occurs between 2 and 3 years.** Reattaches the IVC from the heart to the pulmonary artery. A fenestration is sometimes created to allow blood to flow back to the heart. This prevents damage to the lungs or right-sided volume overload caused by overflow during the postoperative period.[1,3]

Norwood Operation Performed With Deep Hypothermia and Circulatory Arrest Using Modified Blalock–Thomas Taussig Shunt

1. Create a midline sternotomy and enter the pericardium.
2. Cannulation: A single right atrial cannulation with the arterial cannula placed into the pulmonary artery trunk.
3. Place snuggers around brachiocephalic and main pulmonary arteries. (This allows systemic perfusion to take place through the PDA.)
4. Isolate the main pulmonary arteries (if severe aortic stenosis is present, this is done after hyperthermia-induced ventricular fibrillation). Begin cooling over 25 to 30 minutes. Advance the arterial cannula into the PDA and transect the main pulmonary trunk.
5. Isolate pulmonary arteries using a patch. A variety of patches can be used.
6. Once the target temperature is reached (18 °C), the surgeon may proceed with the correction.
7. Atrial septectomy: Approach through localized atriotomy or appendage. Resect the septum primum (*most

often located inferior to the septum secundum). This step allows for intracardiac mixing of blood.
8. Ligate and divide the PDA from the isthmus of the aortic arch, leaving the attachment to the main pulmonary artery intact.
9. Aortopulmonary amalgamation: create an extensive incision in the aorta. Align the ascending aorta with the pulmonary artery and perform a side-to-side anastomosis using a pulmonary homograft and running suture line.
10. Aortopulmonary shunt: Begin warming the patient. Attach a PTFE systemic-to-pulmonary artery shunt (3.0-3.5 mm diameter) from the brachiocephalic artery to the origin of the right pulmonary artery.
11. Close and wean the patient off of bypass.[1,3]

Modified Norwood Operation Using Regional Perfusion Techniques and a Sano Shunt

Most commonly used perfusion method in HLHS.

1. Create a midline sternotomy and enter the pericardium.
2. Cannulation: A single right atrial cannulation with the arterial cannula is placed into the pulmonary artery trunk. Suture a PTFE graft into the brachiocephalic artery for arterial perfusion during the remainder of the operation.
3. Initiate CPB and begin cooling the patient over 25 to 30 minutes.
4. Isolate the main pulmonary arteries: (if severe aortic stenosis is present, this is done after hyperthermia-induced ventricular fibrillation). Advance the arterial cannula into the brachiocephalic artery and transect the main pulmonary trunk. Place a vascular clamp below the origin of the PDA. Isolate the pulmonary artery with a PTFE graft.
5. Initiate cardioplegia: a special 3-way stopcock in the arterial.
6. Aortopulmonary amalgamation: Create an extensive incision in the aorta. Align the ascending aorta with the pulmonary artery and perform a side-to-side anastomosis using a pulmonary homograft and running suture line.
7. Placement of Sano shunt: right ventricle-to-pulmonary artery shunt.

Begin rewarming the patient. Remove and clip the PTFE graft from the pulmonary artery. Slightly enlarge the hole and attach the Sano shunt.

8. Attach the other end of the Sano shunt to the right of the ascending neoaorta.
9. Close and wean the patient off of bypass.[1,3]

Bidirectional Glenn Shunt

Pulmonary artery isolation is most likely done before a Glenn shunt as part of the first stage Norwood operation. If not, these steps are done before the SVC transection.[1,3]

1. Initiate CPB. Crossclamping and cardioplegia are initiated to prevent air embolism if the pulmonary artery is still attached to the heart.
2. Clamp SVC near the insertion of right atria. Transect SVC.
3. Attach and oversew SVC into pulmonary artery using interrupted suture technique and 2 suture lines.
4. Wean off of CPB.[1,3]

Extracardiac Fontan operation with a fenestration.

1. Initiate CPB. Cannulate the neoaorta and apply an aortic crossclamp. Initiate cardioplegia.
2. Clamp right atria across the IVC-atrial junction (*avoid injuring the right coronary artery and tricuspid valve*).
3. Once the clamp is placed and secured, a suture is placed into the IVC. This is done before resection to prevent retraction below the diaphragm.
4. Transect IVC and close right atrium.
5. Remove clamp.
6. Use a PTFE graft to anastomose IVC to the pulmonary artery. Complete attachment using a running suture technique. Attachment site is near previous SVC anastomosis.
7. Wean off of bypass[1,3]

Notes

Complications. Hemodynamic instability and collapse. Requires very careful monitoring postoperatively.

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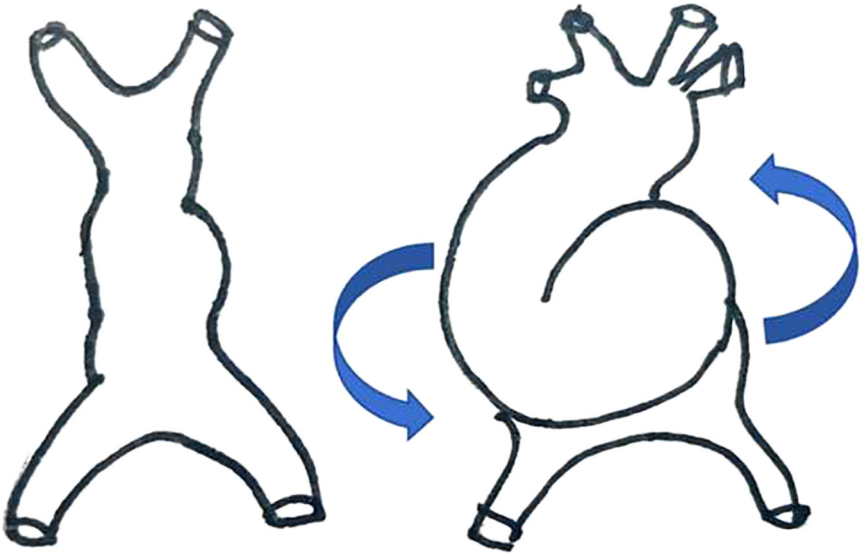

FIGURE E1. Looping of the heart tube during embryological development. Shown inline here for ease of reading.