Abstract:
Children with congenital heart disease represent a unique group of patients presenting to the emergency department. Providing appropriate care to this population requires that the involved medical personnel have an understanding of basic anatomy and physiology both precorrective and postcorrective surgery. Providers must recognize common presentations of illnesses and complications specific to these patients, as well as routine general pediatric illnesses. This chapter covers the basics of congenital heart disease for emergency medical services providers, nurses, and physicians as well as key aspects for managing both routine and complicated illnesses in this population.

Keywords: congenital heart disease; truncus arteriosus; transposition of the great arteries; tricuspid atresia; hypoplastic left heart; tetralogy of Fallot; total anomalous pulmonary venous return; coarctation of the aorta

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Congenital Heart Disease: Complications Before and After Surgical Repair

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VIGNETTE
A parent finds her 18-month-old child with a history of single ventricle and recent congenital heart surgery crying and cyanotic. She is concerned because her daughter has been very irritable, with decreased activity and decreased oral intake. The child has developed a low-grade fever with coughing and episodes of vomiting. Mother is concerned because the patient’s oxygen saturations while sleeping are significantly lower than normal (saturations usually reside in the high 80s). She called emergency medical services (EMS) because the patient has become difficult to arouse.

EMERGENCY MEDICAL SERVICES ASSESSMENT
Children with congenital heart defects can be a challenging group of patients to manage, especially if you are not commonly exposed to these children. Emergency medical services (EMS) personnel should focus their treatment on stabilizing airway, breathing, and circulation (the ABCs) and getting the patient to the emergency department (ED) safely.
One of the most important assessment resources an EMS provider can use is the patient’s caregivers. Children with cardiac defects can often be pale and cyanotic and have increased work of breathing at their baseline. Most parents will be able to help guide assessment and treatment of these children. First, assure that the airway is patent and then assess the child's breathing pattern. Always involve the parents and caregivers in the assessment of the patient's breathing pattern, work of breathing, and overall baseline effort. Questions regarding respiratory status should always be referenced in relationship to the patient's baseline. Another key component of the cardiopulmonary system assessment in patients with corrected congenital heart disease is their baseline oxygen requirement and oxygen saturation. If the parents state the oxygen saturations have been low in comparison with the child's baseline, then apply supplemental oxygen, although start with a small amount of additional oxygen and titrate the supplemental oxygen to meet the child's baseline oxygen saturation goal. Once the baseline has been met, additional supplementation may prove to be harmful. The cardiovascular system in children with congenital heart disease may have altered pathways of blood flow; too much oxygen can cause increased blood flow to the lungs leading to pulmonary edema.

The assessment of circulation in a patient with a cardiac defect includes evaluation of the child's color, capillary refill time, central and peripheral pulses, and blood pressure. If the EMS assessment reveals compromise in the circulatory system, a peripheral intravenous (IV) catheter should be placed. A fluid bolus of normal saline may be given; the dose for children with cardiac defects at 5 to 10 mL/kg. Achieving vascular access in children with cardiac defects is often difficult. If circulatory compromise is present and IV access cannot be obtained, intraosseous access should be considered. This decision should be tempered in relationship to the patient's degree of compromise and the perceived risk-benefit ratio. Other factors that should be considered before IV placement are the skill set of each provider encountering the child. The person who is best at starting IVs should attempt IV placement on these children because crying and fussiness with prolonged access efforts may actually worsen the patient's condition. If there is minimal circulatory compromise noted on evaluation, it may be better to transport the child to a more controlled environment where the IV can be placed.

The last consideration is patient transport. If the child is stable, it may be more beneficial for this patient to come directly to a medical center that has pediatric cardiology specialists. Of course, if the child is unstable, he/she should be taken to the closest medical facility for stabilization. Input from parents and/or experienced caregivers may be helpful in determining the most appropriate receiving facility.

Perhaps the most important take-home point in the prehospital care of children with congenital heart disease is involving the parent or caregiver. Not involving or eliciting input from these resources only increases the risk for inaccurate patient assessment and deterioration due to inappropriate medical interventions. These children are unique in many aspects, and their disease state, adaptability, and response to stress and medical interventions are very patient specific and cannot be generalized. Therefore, using the parents' intuition and knowledge about their child's baseline and their experience with past illness is key toward accurate patient assessment and effective interventions by EMS personnel stabilizing and transporting these patients.

Another important aspect of prehospital care of these children is the transmission of information to the receiving ED. During a medical report, it is important to give a detailed description of the patient's underlying cardiac issues, current vital signs (with baseline vitals if the caregiver was able to provide this), and patient assessment with particular interventions that were required. The call should end with a final assessment of the needs that this particular patient may require from the ED staff. This information from EMS will greatly enhance the quality and efficiency of the care provided in the ED.

**EMERGENCY DEPARTMENT NURSING CARE**

The bedside nurse will often encounter these patients before the doctor is available to see them. Assessing a child with a congenital heart defect should begin just like the assessment of any adult or pediatric patient, ensuring that the ABCs are intact. Remember that the best resource for information regarding this child may be the parents or caregivers as they will help guide your care and be able to answer many questions. The critical thinking process for these patients with corrected congenital heart will be based on a primary assessment by the ED nurse focused on determining if the patient is in respiratory distress or failure, or has evidence of heart failure or cardiovascular compromise.
The nursing assessment for a child with a congenital cardiac defect should include the following:

Airway: is the airway patent or in jeopardy—yes or no?

Breathing: is the patient in respiratory distress or failure—yes or no?

- Are there retractions, nasal flaring, increased respiratory rate, or a decrease in oxygen saturation? Remember that providing supplemental oxygen unnecessarily, or too high of a flow rate or concentration, may change the flow of blood in the heart, leading the child to decompensate. Nurses should consider using a blender for oxygen delivery in these children; if this is unavailable, then starting with a small amount of oxygen and titrating up to the child's goal saturation are acceptable.

- While listening to breath sounds, heart sounds should also be auscultated. Does the child have a murmur, or can you hear the shunt? Murmurs can be hard to auscultate, but hearing a murmur in a child with a Blalock-Taussig shunt is a normal finding. If you are reassessing a patient and the murmur has become softer, make sure that the physician is aware of this change because this could be a sign of shunt compromise.

Circulation: is the patient in heart failure—yes or no?

- Assessment for circulation should include the child's color, capillary refill, central and peripheral pulses, blood pressure, and the palpation of the liver for hepatomegaly. If an infant presents to the ED and there is suspicion of a cardiac defect, blood pressure should be measured on all 4 extremities. Normally, the upper- and lower-extremity pressures are nearly the same, with the lower extremities having slightly higher systolic pressures than the upper extremities. If a difference of greater than 20 mm Hg exists between the upper and lower extremity blood pressures, then it is possible that a coarctation is present.

- Another aspect that needs to be discerned is the following: does the patient have a history of underlying cardiac dysrhythmias at baseline, managed by certain medications? And if so, what interventions in the past have been used to correct dysrhythmias?

During the nursing assessment, the child should be placed on a cardiac monitor, and vascular access obtained, with laboratory studies and fluid resuscitation initiated.

- These patients should always be placed on full cardiac monitoring. The heart rhythm will often not be normal sinus rhythm. It is important to compare the rhythm to previous rhythm strips and electrocardiograms (ECGs) to determine if there are any changes in the patient's cardiac rhythm.

- Peripheral IV placement in a child with a cardiac defect can be a challenging task; therefore, the person with the most skill at placing an IV should be used for this procedure. Multiple IV attempts can cause prolonged crying that can alter the flow of blood, causing oxygen desaturation. If IV attempts are unsuccessful, then an intraosseus needle (IO) should be used.

- Blood should be drawn with the IV start. Useful laboratory tests include a complete blood count and serum electrolytes including magnesium and phosphorus. Other laboratory test results that are commonly obtained include B-type natriuretic peptide (BNP), ionized calcium, prothrombin time, and partial thromboplastin time. Less commonly ordered studies include troponin and creatine phosphokinase MB isoenzyme. If available, a venous blood gas should be obtained to provide a quick reference to the child's metabolic state and electrolyte balance and to determine if the lactate is increased.

- Fluid resuscitation in a child with cardiac defects requires close monitoring and reassessment of the child. Fluid boluses in these children should begin with 5 to 10 mL/kg of isotonic fluid (normal saline). One of the most important aspects of fluid administration in children with single ventricle cardiac defects is to make sure that there are no air bubbles in the tubing as air in the line can cause these children to have a stroke. If the child needs fluids, the bolus may need to run in over a half hour to an hour. Reassessment of breathing and liver size should be done before additional
boluses to make sure that the child is not being put into a fluid overload state.

History: as stated previously, the parents are going to be an invaluable resource for this patient and can help guide your care. The following are good questions to ask the family when assessing your patient:

• What is the child’s normal oxygen saturation?
• How does the child look to them? Ask about normal activity, playfulness, whether the child cries around strangers, and how they are interacting with you.
• What is different today? Is there increased respiratory effort, or did they have to turn the oxygen up at home?
• Is there breathing difficulty while feeding, or does the child sweat or tire easily during feeding?

CYANOTIC CONGENITAL HEART DISEASE: OVERVIEW

Cyanotic congenital heart disease (CHD) encompass all heart lesions present at birth that result in cyanosis, or poor blood oxygen levels. Patients born with these lesions may have been diagnosed in utero by prenatal ultrasound or may go undiagnosed until birth or shortly thereafter. Each lesion presents at a different time based on several factors including (but not limited to) type of defect, presence of associated lesions, severity of the congenital defect(s), and gestational age at the time of delivery. In this section, we will review the most common cyanotic heart lesions (and 1 noncyanotic lesion), go through how they change as they are repaired, and discuss complications specific to the lesion and repair. For some of the lesions, specific management advice is covered within the section. At the end of the section, general management is reviewed.

It is important to remember that despite having the underlying condition, patients with CHD may be presenting with an unrelated general pediatric illness. This illness may or may not be impacted (or have an impact on) their underlying heart disease. For example, a patient with CHD presenting with acute otitis media who is otherwise well hydrated and without respiratory issues may simply require routine antibiotic treatment. In contrast, a patient with CHD presenting with gastroenteritis or bronchiolitis—where dehydration and respiratory function may be complicated or may complicate the underlying heart disease—may need additional support in comparison with the otherwise healthy pediatric patient. It is important for the physician and other ED staff providing care to the patient to be aware of the patient's underlying disease and how it may complicate routine pediatric illnesses. Furthermore, it is important for emergency care providers to decipher if the patient's CHD is playing a role in the patient's current illness and symptoms because the appropriate intervention may need to be adjusted for the patient with CHD.

The 5 “Ts” of cyanotic heart lesions are as follows:

1. Truncus arteriosus
2. Transposition of the great arteries (TGA)
3. Tricuspid atresia, including hypoplastic left heart syndrome (HLHS)
4. Tetralogy of Fallot (TOF)
5. Total anomalous pulmonary venous return (TAPVR)

Truncus Arteriosus

Truncus arteriosus is a lesion where both the aorta and the pulmonary artery are together as a single trunk arising from normally formed ventricles (Figure 1). These children, thus, have mixed blood circulating through their body by way of this common trunk. Before surgical repair, patients with truncus arteriosus may have oxygen saturations in the 70 to 80% range.

Repair is usually done around 2 weeks of age. The pulmonary arteries are taken off of the common trunk and connected to the right ventricle (RV) via a homograft or conduit (Figure 2). The associated ventricular septal defect (VSD) is also closed at this time.

Complications arising from this surgery include valvular insufficiency, arrhythmias, and outgrowth of the conduit.

Figure 1. Truncus arteriosus. (Reprinted with permission of Mayo Foundation for Medical Education and Research. All rights reserved.)
1. With valvular insufficiency, the child may present with pulmonary congestion or pulmonary hypertension crises. Patients in this scenario may benefit from furosemide or other diuretics. You may appreciate crackles on lung examination or respiratory distress in these patients, and a chest x-ray may show pulmonary congestion with increased lung markings. Use caution in giving patients in this scenario oxygen because this may vasodilate the lungs further and worsen the condition.

2. Common arrhythmias seen are right bundle-branch blocks and heart blocks.

3. If the patient has outgrown the conduit, it may not be supplying enough blood to the lungs, and in this instance, patients may benefit from oxygen supplementation.1-5

Transposition of the Great Arteries

This is a lesion where the pulmonary and aortic trunks are placed on the opposite ventricle, thus requiring on the right side of the heart to pump blood to the body (Figure 3). This lesion relies on some sort of mixing lesion in association with the transposition to be compatible with life (atrial septal defect [ASD], VSD, or patent ductus arteriosus [PDA]). There are 3 surgical repair approaches that these patients may have had; the most common is the arterial switch.

1. Arterial switch: in this repair, the pulmonary and aortic trunks are simply switched to the proper ventricle. Although the trunks are switched, the valve stays with its original ventricle. The coronary arteries are also disconnected and then reimplanted onto the new aortic valve (Figure 4).

2. Rastelli procedure: this is used in cases where the patient had left ventricular (LV) outflow tract obstruction and an associated VSD (Figure 5). During the repair, the VSD is closed such that the LV pushes slightly into the RV, thus alleviating the obstruction in outflow. A conduit is then created from the RV to the pulmonary artery.

3. Mustard/Senning procedure: this is rarely seen in practice; however, some cases do still arise (Figure 6). This approach is used when the patient is felt to have poor LV function and, thus, will need to rely on the RV for systemic circulation (or in cases when the coronaries are in an aberrant location). In this repair, the atria are switched (instead of the usual arterial switch). Ultimately, the LV pumps to the lungs and the RV pumps systemically.

There are several complications that may arise from the repair of TGA:

1. Myocardial ischemia: most often seen in cases where coronaries are reimplanted and may have poor/inadequate flow. These patients may benefit from IV fluids, oxygen, and general supportive care.

2. Supravalvar stenosis: seen especially with the Rastelli procedure, where the patient had LV outflow tract obstruction presurgical.
repair. These patients may benefit from IV fluid to overcome the stenosis. They may also benefit from drugs that decrease afterload (nitroprusside), thus allowing the LV to overcome the obstruction with less strain.

3. Arrhythmias: these are more commonly seen with Rastelli and Mustard/Senning repairs, where the RV has been manipulated.

4. Poor LV function: more commonly seen in patients undergoing the Mustard/Senning procedure or in patients where surgical repair was performed later in life.1-3

Tricuspid Atresia and Hypoplastic Left Heart

Although the 2 lesions differ anatomically, functionally they both are considered “single ventricles” and the surgical repairs are almost identical for both. Figure 7 depicts tricuspid atresia in which the tricuspid valve is poorly formed, thus resulting in an underdeveloped RV. Figure 8 depicts HLHS in which the mitral valve is poorly formed, resulting in an underdeveloped LV and atretic aorta.

In both cases, the patient will rely on a mixing lesion (ASD, VSD, PDA) to supply oxygenated blood to the body. Oxygen saturations will be in the 70 to 85% range before surgical correction. Repair of these lesions is done in 3 stages, as described below:

- Norwood procedure (done only in HLHS)
- Stage I: Blalock-Taussig shunt (BTS) or Sano modification (at birth)
- Stage II: Glenn shunt (6-9 months)
- Stage III: Fontan procedure (18-36 months)
1. Norwood + BTS or Norwood + Sano (Figure 9)
   - The Norwood procedure is only done in HLHS as these children have an atretic aorta. This procedure creates a new (neo) aorta from the existing aorta and part of the pulmonary artery. The main pulmonary artery is then ligated.
   - BTS is the creation of a conduit from the right subclavian artery to the right pulmonary artery, thus allowing for pulmonary blood flow.
   - Sano modification is a creation of a conduit from the RV to the pulmonary artery.
   - In all cases, an ASD will be created or augmented to allow mixing of blood returning to the heart. By the end of this first stage of surgical repairs, the patient will be considered a “single ventricle.”

- The decision to use a BTS or Sano modification is based on the patient and the surgeon. Both procedures are used until the second stage of repairs can be completed. Each has its advantages and disadvantages.
  - BTS does not cut into the RV; however, during diastole, the BTS will “steal” blood from the coronaries, thus resulting in poorer coronary blood flow.
  - The Sano requires cutting into the RV, which may result in arrhythmias and abnormal conduction. However, it allows for improved coronary blood flow.4,5

Figure 6. Mustard/Senning repair for transposition of the great arteries. (Reprinted with permission from www.med.umich.edu. ©Regents of the University of Michigan, 2012.)

Figure 7. Tricuspid atresia. (Reprinted with permission of Mayo Foundation for Medical Education and Research. All rights reserved.)

Figure 8. Hypoplastic left heart syndrome. (Reprinted with permission of Mayo Foundation for Medical Education and Research. All rights reserved.)
Figures 9. A, Norwood procedure and BT shunt. B, The Sano modification. (Fig 9A reprinted with permission of the Mayo Foundation for Medical Education and Research. All rights reserved. Fig 9B reprinted with permission from Children’s Hospital Boston, http://www.childrenshospital.org/cfapps/mml/index.cfm?CAT-media&MEDIA_ID=2024)
Oxygen saturations after this procedure are generally 75 to 85%.

2. Glenn (Hemi-Fontan) procedure (Figure 10)
- The BTS or Sano is taken down. The superior vena cava (SVC) is then connected either to the right pulmonary artery or to the main pulmonary artery (bidirectional Glenn). This results in passive blood flow directly into the lungs from the upper body. Blood returning from the lower body continues to flow into the right atria. Saturations after this procedure are generally 75 to 85%.

3. Fontan procedure (Figure 11)
- This is the final stage of the repair where the inferior vena cava (IVC) is connected to the pulmonary arteries, thus allowing all blood flowing back to the heart to flow directly (passively, because no valve exists) into the lungs, and thus, bypassing the right atrium (RA). This leaves the heart responsible solely for pumping oxygen-rich blood to the body. Once complete, patients usually have oxygen saturations in the normal reference range. This procedure may be accomplished in a few different fashions:
Extracardiac: the IVC is connected to the pulmonary artery (PA) without being connected to the heart at all.

Intracardiac: the IVC is connected to the heart by way of “tunneling” through the RA.

In either type, a fenestration may be placed at the RA serving as a “pop-off valve” in cases where pulmonary resistance is high and blood is backing up.¹⁻³,⁶

Complications arising from this 3-stage repair are several and are associated with the manipulations being performed.

1. When a shunt is placed, the size is very important. Too small of a shunt will prevent adequate flow and lead to backup, whereas a large shunt will lead to fluid overload (in a BTS, this will result in pulmonary edema).⁶

2. In connecting the SVC and IVC directly to the PA, no valve or pumping device exists in the situation where pulmonary resistance increases. Thus, it is impossible to overcome this resistance, and blood flow will back up, resulting in SVC syndrome and/or liver enlargement.

3. If a fenestration is placed for a “pop-off” in cases where pulmonary vascular resistance (PVR) is elevated so that blood flow does not back up significantly, emboli may occur.¹,⁵

Tetralogy of Fallot

Tetralogy of Fallot consists of 4 components (Figure 12):

1. Pulmonary infundibular stenosis
2. Overriding aorta
3. Right ventricular hypertrophy
4. Ventricular septal defect

The component with the most significant impact on the severity of the condition is the pulmonary stenosis. Thus, children with more profound pulmonary stenosis are often more symptomatic and may require earlier correction (or palliation until correction).

TET Spells

TET spells, seen specifically in patients with uncorrected TOF, occur when there is an increase in right to left shunting. The cause of the increase in shunting is any increase in lung pressures (including crying), which leads to higher pulmonary artery pressures and, thus, more blood flow shunting across the VSD down the path of least resistance. The result is an increased amount of oxygen-poor blood-entering circulation. Treatment for this condition is based on increasing systemic vascular resistance (SVR), thus decreasing the right to left shunting. All of the options below may be used alone or in conjunction with each other to increase SVR and decrease pulmonary pressures:

1. Place the child in a knee to chest position
2. Try and calm the child (stop crying)
3. IV fluids
4. Oxygen
5. Morphine

Repair for patients with TOF is often performed around 6 months of age. The procedure entails closing the VSD and augmentation of the pulmonary artery. In severe cases, a conduit may need to be created to bypass the pulmonary stenosis. Complications that may arise secondary to this repair include the following:

1. Residual VSD
2. RV failure—this may be secondary to significant hypertrophy before surgical correction or in cases of significant residual VSDs
3. Conduction abnormalities—this is more common in cases where a conduit was created and the RV wall was surgically manipulated
4. Valvular insufficiency—seen more often when a pulmonary conduit is placed¹⁻³
Total Anomalous Pulmonary Venous Return

Patients with TAPVR have pulmonary veins that connect to systemic (non-pulmonary) veins, and finally, draining into the RA (Figure 13). Thus, the RA receives both oxygenated and deoxygenated blood. For survival, these patients must have some mixing lesion (usually an ASD; sometimes a VSD). There are several courses the veins may take to ultimately reach the RA, and the course is a significant component in determining the severity of the disease.

The 4 main types of TAPVR are as follows:

1. Supracardiac: pulmonary veins drain into the RA via the SVC
2. Infracardiac: pulmonary veins drain into the RA via hepatic veins and the IVC
3. Cardiac: pulmonary veins drain into coronary sinus
4. Mixed: pulmonary veins split-up and drain via more than 1 of the above options

The repair of TAPVR involves detachment of the pulmonary veins from the RA and reattachment to the LA. The intracardiac mixing lesion is closed. Complications from this repair are generally related to stenosis of the anastomosis resulting in pulmonary edema. In these cases, decreasing SVR or short-term use of diuretic may be useful to prevent pulmonary overload.1-3
Acyanotic Lesions—Coarctation of the Aorta

In this condition, there is a narrowing of the aortic arch, most commonly just beyond the aortic arch in the “juxtaductal region” because the narrowing is often near or at the site of the PDA (Figure 14). The degree of narrowing, along with any associated lesions (PDA, VSD, bicuspid aortic valve), determine the timing and severity of presentation. Symptoms may be nonspecific, including tachypnea, tachycardia, poor feeding, and vomiting. Other, more specific findings include congestive heart failure (especially if an ASD/VSD is present), lower-extremity cyanosis, poor femoral pulses, lower blood pressure, and oxygen saturations in the lower extremities. A murmur may or may not be present. In patients who have been repaired, the most common complication is recurrence of the coarctation.

ASSESSMENT AND MANAGEMENT OF ILL CHILDREN WITH CONGENITAL HEART DISEASE

The assessment of the child with known or suspected CHD—whether repaired or unrepaired—should follow a systematic approach (see Figure 15). The emergency management of these children should encompass both general interventions and those specific to the suspected underlying cause.

General Interventions

A general approach to the child with CHD should include the following considerations:

1. ABCs
2. Consider supplemental oxygen
   a. Is this patient below their baseline oxygen saturation? If so, attempt to administer oxygen to get them back to their baseline (not to 100%—unless that is their baseline).
3. Vascular access
   a. Caution with line placement in the neck as resultant clots may prevent patient from future operations (Glenn and Fontan).
4. Prudent fluid management
   a. This can start with a 5 to 10 mL/kg bolus if unsure of fluid status.
   b. If concern exists for dehydration leading to thrombus and/or other complications, consider 10 to 20 mL/kg bolus.
5. Laboratory studies: venous blood gas, complete blood count, complete metabolic panel, and B-type natriuretic peptide
6. Chest radiograph—2 views
7. ECG
   a. Look for changes from previous ECGs, including rhythm changes and new hypertrophy/strain patterns.
8. Ultrasound to assess heart function and fluid around the heart
9. Cardiology consult
10. Echocardiogram

Disease-Specific Interventions

1. Suspected shunt malfunction
   a. If a shunt is too large, increased pulmonary flow and overload may occur, especially in cases of RV-PA conduits and BTS. In these patients, decreasing blood flow to the lungs is key; thus, decreasing inspired oxygen to vasoconstrict pulmonary flow may be useful along with diuretics. Look for signs of pulmonary congestion and respiratory distress.
   b. If the shunt is too small, just the opposite may occur, with too little flow reaching the lungs. In these patients, oxygen administration to vasodilate the lungs may be beneficial. Look for signs of jugular venous distention and for liver enlargement without evidence of pulmonary congestion. These patients are likely to have increased cyanosis.
   c. Thrombus formation within the shunt occurs more often in states of dehydration. In these patients, appropriate fluid resuscitation is key. Oxygen administration may be useful, but proceed with caution and monitor respiratory status closely. These patients will typically have had an acute decompensation and drop in oxygen saturation, usually in a state of dehydration.  

2. Fenestration
   a. Although helpful in providing a pop-off valve in states of high pressure with difficult flow, fenestrations allow for the formation of emboli. Treatment is based on where the emboli are located. Look for a patient with an acute decompensation/sudden change in neurologic status.

3. RV conduit complications
   a. Arrhythmias—including bundle-branch blocks. Usually, these are noted immediately postoperatively, and proper precautions are taken to maintain adequate function (eg, via pacemaker). However, if unrecognized postoperatively, or in an event where a pacer is not working properly, patients may require medications or external pacing until a definitive solution can occur. These patients may present with episodes of syncope or be called in because of a known malfunction (ie, malfunction was noted during a recent interrogation).
   b. Over time with scarring and reorganization of the RV, function may decrease resulting in right heart failure, leading to signs of jugular venous distention and liver enlargement. Medical therapy may be necessary to
help these patients through acute issues until long-term therapy can be determined.

4. Complications of improved pulmonary circulation
   a. With the benefit of improved pulmonary circulation comes the risk of pulmonary overload, leading to edema, effusions, and backup. These patients will often require surgical correction, but in the acute setting, management with diuretics and careful oxygen control to prevent further overload are key.

5. Complications of medical therapy to prevent fluid overload
   a. Medical therapy is integral to many patients to control fluid status and prevent fluid overload. However, when these medications work too well, or in cases where the patient becomes dehydrated secondary to acute illness, poor flow, and possibly, thrombus formation may occur.

These patients need careful fluid resuscitation with 10 to 20 mL/kg bolus(es). Look for a patient with an acute change in cardiopulmonary status with an intercurrent illness.

**Cardiogenic Shock**

The patient assessment findings that you would see with cardiogenic shock are the same as other shock etiologies. This includes weak pulses, poor capillary refill time, decrease in urine output, change in mental status, and hypotension. Specific assessment findings for cardiogenic shock include cardiac dysrhythmias, S3 or S4 heart sounds, jugular venous distension (difficult to assess in young children), edema, and hepatomegaly. Aggressive fluid volume replacement is generally needed for the shock state. In patients with CHD, although caution should be taken with fluid resuscitation, judicious fluid boluses (20 mL/kg) with reassessment after each bolus may be indicated to overcome...

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<td>Sildenafil</td>
<td>PDE I</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>PO/NG start at 0.25 mg/kg Q6 and tiritate</td>
<td>hypotension</td>
</tr>
<tr>
<td>Nitroprusside</td>
<td>Vasodilator A-V (↓ afterload)</td>
<td></td>
<td></td>
<td>dilatation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vasopressin</td>
<td>AVP1 receptor</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0.003-0.002 units/kg/min</td>
<td>bradycardia, hypotension</td>
</tr>
<tr>
<td>Amiodarone</td>
<td>Class III antiarrhythmic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>load 5 mcg/kg; Infusion 5-10 mcg/kg/min</td>
<td>slows repolarization and AV conduction; prolongs QTc</td>
</tr>
</tbody>
</table>

**Figure 16.** Cardiovascular drugs and drips. Abbreviations: A, arterial; B, beta; PDE I, phosphodiesterase inhibitor; PO, by mouth; NG, nasogastric; V, venous; AVP1, arginine vasopressin 1.
the patient's hypovolemia. If during the fluid resuscitation, auscultation of the lungs reveals crackles and respiratory distress ensues or worsens, a diuretic should be considered.7

If the patient continues to have poor perfusion and hypotension despite adequate fluid administration, then an inotrope and vasopressor may be needed to support perfusion and blood pressure. Dopamine (0.5-20 μg/kg/min) is frequently the first-line inotrope used for cardiogenic shock. Often, Milrinone (0.25-1 μg/kg/min) is chosen because it reduces afterload and has a positive inotropic effect on the heart. Dobutamine, epinephrine, and nor-epinephrine can all be used for their inotropic effects (see Figure 16 for dosing).7,8

**Intubation, Ventilator Set-up, and Cardiopulmonary Resuscitation in Congenital Heart Disease**

In cases where the patient must be intubated, maintaining the balance of adequate ventilation and oxygenation along with pulmonary and systemic perfusion can be challenging. Positive pressure ventilation of any sort in patients with cyanotic heart lesions can lead to decreased pulmonary blood flow as the positive pressure impedes pulmonary flow. Specifically, patients with Glenn or Fontan physiology are more likely to have increased PVR, leading to decreased cardiac output. They lack an RV response to overcome this PVR because the Glenn and Fontan allow passive flow from the SVC and IVC, respectively. In patients with TOF with RV hypertrophy and RV dysfunction (which may be seen after surgical repair), diastolic filling is impaired, resulting in poor cardiac output (Figure 17).9

Therefore, if intubation or positive pressure ventilation of any sort is necessary, special considerations must be made to achieve this balance. Blood pressure, perfusion, and respiratory status must be carefully monitored to ensure that this balance is met and maintained. Low end-expiratory pressure is essential to allow for adequate blood

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Repair</th>
<th>Complications</th>
<th>Oxygen Saturations</th>
</tr>
</thead>
</table>
| Truncus Arteriosus            | 1. Pulmonary arteries moved from the main trunk and connected to RV  
                               | 2. Patch closure of VSD                                                      | Pre-repair: mixing lesion - sats 75-85%   |
|                               |                                                 | 1. Arrhythmias  
                               | After repair: ~100%                                                        |
|                               |                                                 | 2. Valve insufficiency leading to pulmonary hypertension and pulmonary edema |                                           |
| Transposition of the Great Arteries | Arterial switch (back to normal position) or Rastelli (uses a conduit from RV to PA) or Mustard/Senning (Atrial Switch) | 1. Myocardial ischemia  
                               | Pre-repair: mixing lesion - sats 75-85%                                    |
|                               |                                                 | 2. Arrhythmias  
                               | After repair: ~100%                                                        |
|                               |                                                 | 3. Valvular stenosis  
                               |                                           |
|                               |                                                 | 4. Poor LV function and CO                                                  |                                           |
| Tricuspid Atresia Hypoplastic Left Heart Syndrome (HLHS) | 1. Norwood +BTS or Sano (birth)  
                               | 2. Glenn (6-9 months)  
                               | 3. Fontan (18-36 months)                                                    | Pre-repair, post Norwood and Glenn: 75-85% |
|                               |                                                 | 1. Norwood BTS or Sano: shunt thrombosis, pulmonary edema or cyanosis if shunt size not correct  
                               | After Fontan: near 100% (Unless patient is utilizing the fenestration)      |
|                               |                                                 | 2. Glenn: SVC syndrome because of increased PVR  
                               |                                           |
|                               |                                                 | 3. Fontan: SVC syndrome if elevated PVR; Risk of emboli if fenestration is placed |                                           |
| Tetralogy of Fallot           | Around 6 months of age:  
                               | 1. Augmentation of PA and valve (or placement of a conduit)  
                               | 2. Closure of VSD                                                          | Pre-repair: dependent on pulmonary stenosis |
|                               |                                                 | 1. Arrhythmias  
                               | After repair: 100%                                                         |
|                               |                                                 | 2. Valve insufficiency leading to pulmonary hypertension and pulmonary edema |                                           |
|                               |                                                 | 3. Residual VSD  
                               |                                           |
|                               |                                                 | 4. RV failure                                                              |                                           |
| Total Anomalous Pulmonary Venous Return | 1. Reattachment of pulmonary veins to LA  
                               | 2. Rerouting (if needed) of pulmonary vessels                               | Anastomosis stenosis with resultant pulmonary edema  |

Figure 17. General overview of congenital heart lesions.
flow. Constant pressure via square wave ventilation is usually recommended. Settings starting with low respiratory rates, short inspiratory time, and a tidal volume of 5 to 6 mL/kg are generally ideal. In cases where a significant pulmonary process may be present, higher inspiratory pressures may be needed initially and can be adjusted as the respiratory status starts to improve.8,9

When setting oxygen levels during ventilation, remember to maintain the patient's baseline oxygen saturations. Remember that oxygen is a vasodilator. It can lead to pulmonary fluid overload in patients not accustomed to such high levels. Conversely, oxygen may be helpful for patients with pulmonary hypertension or poor pulmonary flow.

Cardiopulmonary resuscitation (CPR), likewise, offers physiologic challenges in patients with CHD. This specifically applies to patients like those discussed previously, including those with Glenn and Fontan repairs and TOF physiology. In these patients, CPR, much like mechanical ventilation, creates positive pressure that can impede pulmonary, and ultimately, systemic blood flow.9 However, in situations where CPR is indicated, there is no alternative because it is a potentially lifesaving measure that must be performed. In these scenarios, everything must be done to attempt to alleviate any PVR and restore systemic perfusion.

In patients with CHD (corrected or uncorrected) who require CPR, the importance of coordinated chest compressions with ventilations to assure effective ventricle filling is the same as with other patients, although with a few unique caveats. The use of sodium bicarbonate and calcium chloride/glucagon has been well documented as a beneficial adjunct in these patients during CPR and post-resuscitation stabilization. In these particular patients, significant metabolic acidosis is an underlying issue in relation to the effectiveness of the resuscitation efforts. A state of acidosis is counterproductive to the myocardial function and its responsiveness to resuscitative agents. These patients with cardiac defects are even more affected, and thus, have a less responsive myocardium to resuscitation measures in an acidic environment.7,9 Thus, correction of the underlying acidosis is imperative to improve outcomes.

### SUMMARY

Children with congenital heart disease represent a population of patients presenting to the ED with unique medical issues. It is important for emergency care providers of all levels caring for these patients to understand their underlying heart lesion, repairs, and potential complications specific to these patients. Providers must be aware of how patients with congenital heart disease present with both complications specific to their condition as well as common general pediatric illnesses. Actively seeking baseline information and involving parents and home health care providers are essential in providing the best care to these patients. Furthermore, by remembering the basic principles of congenital heart disease, evaluation and management of these patients will be less intimidating and overwhelming.

### REFERENCES