Chapter 20

Shone Complex

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Case Scenario

A 4-year-old female with a history of Shone complex is scheduled for an umbilical hernia repair. She has a twin sister and is a bit slower and smaller than her twin, requiring more frequent breaks while playing. She has no other medical conditions or comorbidities. Her only previous surgery was a repair of coarctation of the aorta during infancy and she experienced no anesthetic issues. The patient's mother is very concerned about separation anxiety and the effects of anesthesia on her daughter's heart.

On physical exam she appears anxious, hiding in her mother's arms and making no eye contact with healthcare providers. Lungs are clear to auscultation and a 2/6 systolic ejection murmur is appreciated, which is unchanged from prior exams. Vital signs include a heart rate of 98 beats/minute, lower extremity noninvasive blood pressure reading 88/60 mm Hg, respiratory rate 26 breaths/minute, and SpO_2 99% on room air. She appears healthy, with no pallor, cyanosis, or diaphoresis.

Two months earlier, echocardiography showed the following:

- *Parachute mitral valve with mild stenosis, mean gradient 5 mm Hg*
- *Bicuspid aortic valve with moderate stenosis, mean gradient 30 mm Hg*
- *Mild to moderate gradient (10 mm Hg) across the area of aortic coarctation repair*
- *Normal left ventricular function with mild hypertrophy*
- *Normal right-sided structures and function*

Key Objectives

- Describe the characteristic cardiac anomalies of Shone complex.
- Understand treatment options for patients with Shone complex.
- Describe preoperative planning for patients with Shone complex.
- Describe perioperative management for the various cardiac anomalies associated with Shone complex.
- Understand concerns for postoperative pain control and disposition in the Shone complex patient.

Pathophysiology

What is Shone complex?

Shone complex is a group of typically obstructive left-sided lesions of the heart, also referred to as Shone syndrome, disorder, or anomaly. It is a rare congenital cardiac disease, occurring in fewer than 1% of patients with congenital heart disease (CHD). Shone complex was first described by John D. Shone et al. in 1963 as four left-sided heart lesions consisting of a supravalvular ring of the left atrium, a "parachute" mitral valve, muscular or membranous subaortic stenosis, and coarctation of the aorta [1]. (See Figure 20.1.) These anomalies cause a progressive problem with inflow into the left ventricle (LV) and outflow obstruction from the LV and aorta. There is also an association with smaller LV size and decreased LV function. Lesions may progressively worsen over time, causing significant heart failure symptoms, pulmonary hypertension, and arrhythmias [2]. (See Figure 20.2.)

Partial Shone complex comprises two or three of the described four lesions [3]. There may be up to eight lesions (all involving the left side of the heart) and patients may have a combination of several of the lesions. The patient's symptoms are typically related to the lesion that is most severe, with mitral valve stenosis commonly being most symptomatic.

Is there a surgery or technique to correct Shone complex?

The treatment of Shone complex is based on the presence of the lesions and their severity. Each lesion brings a different and distinct set of physiological aberrations. These may act together to cause a series of obstructions that may magnify a single lesion. Each cardiac lesion

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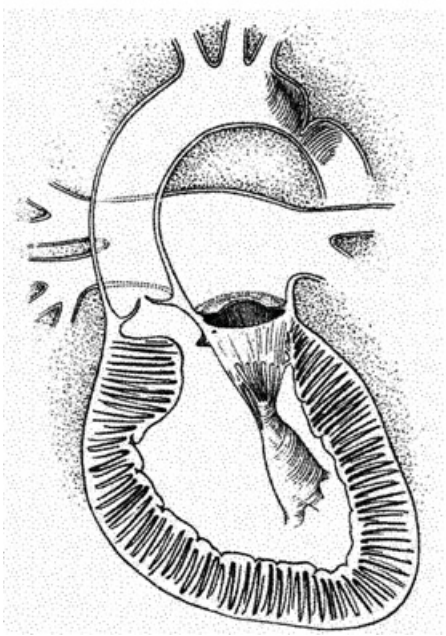

Figure 20.1 Shone complex. Diagrammatic representation of the four obstructive anomalies forming the complex: supravalvular ring of the left atrium, parachute mitral valve, subaortic stenosis, and coarctation of aorta, in that order, according to the direction of blood flow. From Shone J., et al. The developmental complex of "parachute mitral valve," supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta. *Am J Cardiol* 1963; **11**: 714–25. With permission.

must be evaluated individually and then reexamined as part of the whole cardiac complex to understand the pathophysiology and determine management. Each patient with Shone complex has a range in the variety and severity of left-sided lesions and must be evaluated individually: no two patients with Shone complex are the same. There is not a single surgical procedure to correct Shone complex. Surgical intervention may range from a single intervention such as aortic arch repair to a combination of multiple procedures such as repair of aortic coarctation along with mitral valve repair and aortic valvotomy. In severe cases patients may require multiple surgeries for recurrent left-sided obstructive lesions, including mitral and/or aortic valve replacements. (See Figure 20.3.)

Clinical Pearl

No two patients with Shone complex have exactly the same cardiac abnormalities. It is imperative to evaluate and understand each patient's pathophysiology. Surgical intervention may range from a single intervention such as aortic arch repair to a combination of multiple procedures such as repair of aortic coarctation along with mitral valve repair and aortic valvotomy.

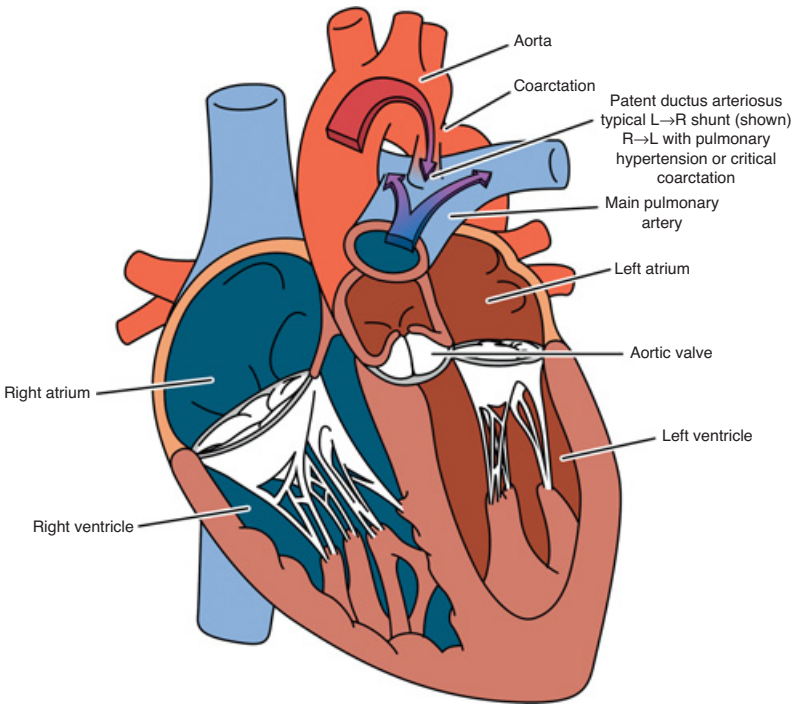

Figure 20.2 Shone complex. Drawing by Ryan Moore, MD, and Matt Nelson.

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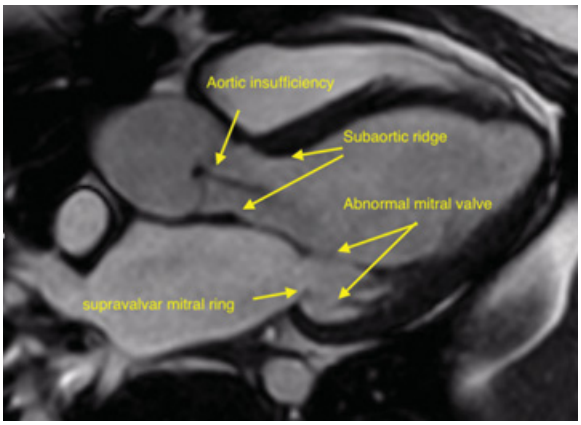

Figure 20.3 Multiple left-sided lesions. Three-chamber magnetic resonance imaging showing supravalvular mitral ring, hypoplastic mitral valve annulus, abnormal mitral valve, small subaortic ridge, and aortic regurgitation. Courtesy of Michael Taylor, MD.

What is a parachute mitral valve?

A normal mitral valve has two leaflets and two papillary muscles; the chordae from each leaflet each insert into a separate papillary muscle. A parachute mitral valve is an abnormality in the mitral valve apparatus in which the chordae of the mitral valve insert into a single papillary muscle [4]. During the first trimester of development, a disruption in embryonic formation of the papillary muscles leads to a single papillary muscle being formed [5]. The chordae in a parachute mitral valve become shorter and thicker, resulting in limited opening of the valve and functional mitral stenosis (MS) [1].

Mitral stenosis, or any LV inflow obstruction, should be evaluated for its etiology and severity. Moderate to severe MS due to a parachute mitral valve may necessitate valve repair or a valve replacement depending on the severity of the stenosis. Congenital MS due to a parachute mitral valve historically has carried a poor surgical prognosis [6]. A supravalvular membrane causing mitral (MS) or aortic stenosis (AS) may require resection depending on how much inflow and outflow obstruction it causes.

Some patients with mitral and aortic stenosis may also have an underdeveloped or hypoplastic LV. The degree of this hypoplasia, LV function, and the ability of the LV to produce adequate cardiac output may all impact surgical planning. A severely hypoplastic LV with a stenotic aortic valve may require single-ventricle palliation. This decision would be made in a multidisciplinary session including cardiologists and cardiac surgeons.

Clinical Pearl

The severity of MS is directly related to the morbidity and mortality of patients with Shone complex. Patients with severe MS are the most symptomatic and have the highest morbidity with surgery.

Why should coarctation of the aorta be corrected?

Coarctation of the aorta causes an increase in LV afterload, hypertension in the head vessels and upper extremities, and hypotension with decreased perfusion to the remainder of the body distal to the narrowing. Without repair, over time, the LV will become hypertrophied and there is an increased risk of coronary artery disease and stroke [7].

What are the surgical approaches to correction of aortic coarctation?

A coarctation repair is generally performed during infancy via a thoracotomy. There are many different approaches to surgical repair such as an end-to-end anastomosis (with a possible extended resection), a subclavian flap angioplasty, and an interposition graft repair. With a subclavian flap angioplasty repair, the subclavian artery becomes discontinuous with the aorta, and consequently blood pressure monitoring on the left upper extremity may not be indicative of the true blood pressure.

Coarctation relief may also be attempted in the cardiac catheterization laboratory via an endovascular approach. Arterial access is obtained through the femoral artery. In infants and small children, angioplasty is typically performed with balloon dilation of the stenosis. In older patients and for re-coarctation of the aorta, angioplasty with stent placement may be performed. Long-term surveillance is required following coarctation repair to evaluate for recurrent coarctation and aneurysm formation. If there is a longer segment of arch hypoplasia, an aortic arch reconstruction may have to be performed via a median sternotomy and utilizing cardiopulmonary bypass.

Clinical Pearl

If the technique previously employed for coarctation repair is unknown, place the blood pressure cuff on the right upper extremity, as this will provide a reliable blood pressure for coronary artery and cerebral perfusion.

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Are Shone complex patients prone to respiratory involvement?

Patients with left-sided obstructive lesions may have increased pulmonary pressures from increased left atrial pressures. Not surprisingly, pulmonary hypertension causes long-term increases in morbidity and mortality in patients with Shone complex [8]. These patients may experience shortness of breath from pulmonary edema. They may also experience an exacerbation of respiratory symptoms with a concomitant respiratory infection.

Clinical Pearl

Patients with Shone complex who have developed pulmonary hypertension have increased anesthetic risk.

Anesthetic Implications

Is a visit to the cardiologist needed prior to elective surgery?

Since the manifestations of Shone complex are so variable, knowing the individual patient's cardiac pathophysiology is of utmost importance. Results of a recent visit to his or her cardiologist should be documented along with an echocardiogram and electrocardiogram (ECG) to delineate disease progression. A discussion with the cardiologist may be beneficial to understand which lesions are causing the most influence on cardiac output and how changes in preload or afterload would affect the patient's hemodynamics. The cardiologist can also help determine if the patient is optimized for an elective surgery.

Once the cardiac pathophysiology is confirmed, an understanding of the patient's functional status and symptomatology is important. Symptoms may occur at rest or with activity. The most commonly elicited symptom is shortness of breath with exertion. Patients with moderate to severe obstructive lesions may have chest pain, dizziness, or even syncope from decreased cardiac output.

How should the type of anesthetic be determined and how should parental concerns about the effect of anesthesia be addressed?

Children with CHD have a disproportionately high risk of perioperative cardiac arrest and anesthetic morbidity [9]. The type of surgery will dictate the anesthetic requirements. For instance, if this patient were to undergo a diagnostic procedure such as magnetic resonance imaging (MRI), sedation or monitored anesthesia care might be appropriate. For an umbilical hernia repair a general anesthetic is required. Maintenance of adequate preload, afterload, and contractility is of utmost importance, along with judicious titration of anesthetic medications.

How could the mother's concerns about her daughter's anxiety be addressed?

Preoperative management is similar to the management of any pediatric patient who will undergo general anesthesia for an umbilical hernia repair. The goal is to minimize the stress response associated with the anxiety of undergoing anesthesia and a surgical procedure. Multiple methods can be used, such as behavioral management techniques facilitated though a child life specialist, pharmacological management with medications such as oral midazolam, and involving a parent in the induction of anesthesia to avoid separation anxiety. The technique or combination of techniques utilized will be determined by the anesthesia team, with the aim of reducing the child's stress while introducing the lowest possible risk. A stress response will elicit tachycardia, which in a patient with Shone complex may cause a reduction in cardiac output and poor perfusion and should be avoided. Child Life specialists, if available, should be utilized early in the preoperative process. Some institutions have a separate preoperative appointment with the Child Life team. Having the child comfortable in his or her surroundings prior to the presence of medical personnel will help reduce the anxiety of a new environment. The use of premedication and/or parental presence depends on the anesthesiologist's comfort with having a parent in the operating room during induction as well as the institutional policy. The safety of the child during anesthetic induction is of utmost importance independent of which modality is utilized to relieve preoperative anxiety.

Should an intravenous catheter be placed preoperatively?

Depending on the severity of the patient's individual pathophysiology induction of general anesthesia may occur either via inhalational mask induction or intravenous (IV) induction. Most patients with mild to moderate MS or AS will be able to tolerate an inhalational induction with vigilant hemodynamic monitoring. Patients with regurgitant lesions will generally tolerate a gentle inhalational induction. This patient has left-sided outflow tract gradients at the mitral and aortic level, as well as a residual gradient at the site of the coarctation repair. Although none of the gradients are severe individually, they should be considered cumulatively and thus a cautious approach to anesthetic induction is warranted. In the patient with

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severe stenosis, it may be prudent to place an IV prior to induction of anesthesia and slowly induce anesthesia with precise hemodynamic monitoring. The specific pharmacologic agents chosen may vary as long as meticulous care is taken to maintaining preload, contractility, and afterload while avoiding the development of significant tachycardia.

Clinical Pearl

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What intraoperative considerations exist with umbilical hernia closure?

Most small umbilical hernias close spontaneously, prior to the child's third or fourth birthday. When the defect has failed to close on its own, is large, or the bowel is at risk for strangulation, the patient will be scheduled for surgical closure. Typically, repair involves a periumbilical incision with primary suture closure of the hernia defect. Unlike the adult population, mesh and laparoscopy is rarely utilized. Depending on the size of the umbilical hernia and the surgeons' approach to the repair, the airway may be secured with either an endotracheal tube or a supraglottic airway device such as a laryngeal mask airway. Some surgeons may also prefer the use of neuromuscular blockade depending on the size of the defect. Spontaneous ventilation, if possible, offers the benefit of having less effect on preload. However, care should be taken to avoid the potentially negative effects of hypercarbia on the pulmonary vascular bed.

How should adequate preload be ensured and monitored perioperatively?

An initial assessment should be made to evaluate the patient's baseline hydration/preload status. Ensure that the patient is appropriately fasted per the American Society of Anesthesiologists (ASA) guidelines but avoid extended periods of fasting that can cause decreased preload. These patients should be scheduled early in the day and should be encouraged to drink clear fluids until 2 hours prior to the procedure. Dehydration should be avoided. If the patient appears dehydrated, has decreased urine output, a lack of wet diapers, or decreased capillary refill, the patient may need to receive IV fluids prior to the induction of anesthesia. Monitoring of preload for a hernia repair is usually not done invasively through a central venous line but is based on clinical signs and symptoms.

Both inhaled and IV anesthetic agents can decrease preload. While one specific anesthetic agent is not preferred over another, the judicious and careful titration of anesthetic agents to the desired effect is recommended.

Management of positive pressure ventilation can also influence preload. While one particular ventilation strategy is not favored over another, maintaining thoracic volumes that allow for optimal venous return to the right side of the heart is important.

The surgical approach can also influence preload. An open repair may manually compress a discrete area of the abdomen whereas a laparoscopic approach may cause unintended compression of small vessels, thus causing decreases in preload and cardiac output. Insufflation pressures can also lead to increased abdominal pressure, resulting in increased intrathoracic pressures and hence an increase in afterload. Maintaining lower insufflation pressures may be necessary when utilizing a laparoscopic approach.

Clinical Pearl

Preload may be affected by hydration status, anesthetic agents, ventilation strategy, and surgical technique.

How should adequate afterload be monitored and ensured?

Afterload is commonly defined as the pressure the LV must produce to pump the volume out of its chamber. It is influenced by the volume in the LV, wall thickness of the ventricle, and systemic vascular resistance (SVR). Patients with Shone complex can have perturbations in all three of these categories.

For the patient with MS, obstruction of blood flow into the LV exists, thus decreasing the volume within the chamber. The LV volume may also be influenced by the patient's heart rate, which relates to the length of time in the cardiac cycle for diastole, or cardiac filling.

Aortic stenosis causes afterload obstruction for the LV. Over time, this increased afterload may result in left ventricular hypertrophy (LVH). An increase in wall thickness may lead to increased LV pressures and decreased or dyskinetic LV function. Maintaining adequate coronary perfusion is critical for a thickened ventricle. Decreases in afterload, commonly seen with induction of anesthesia,

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can lead to decreased diastolic flow to the coronary arteries and myocardium, thus causing ischemia.

From an anesthesia perspective, SVR is the afterload that the LV must pump against. Most anesthetic agents cause a decrease in SVR. Patients with Shone complex may have other perturbations in anatomy that influence SVR such as a coarctation of the aorta and/or AS. Depending on the location of the lesion, there may be an area of "fixed" or relatively high SVR proximal to the lesion and an area of decreased SVR distal to the lesion. The patient with moderate AS has a fixed lesion that is not influenced by the choice of anesthetic. With induction of anesthesia, the LV continues to pump against the same afterload, the stenotic aortic valve. Distal to the aortic valve, if anesthetic agents have caused SVR to decrease, consequently blood pressure and the amount of blood flow to the coronary arteries are decreased. This decrease in coronary perfusion leads to myocardial ischemia, as the LV is required to do the same amount of work to eject blood across the stenotic aortic valve. In the patient with aortic coarctation, the aortic pressure proximal to the coarctation (from the heart to the coarctation, typically around the level of the left subclavian artery) will be higher than the pressure distal to this lesion. This would clinically manifest as a higher noninvasive blood pressure (NIBP) measurement on the right upper extremity compared to the lower extremity.

How should adequate contractility be monitored and ensured?

Contractility is described as systolic function. Echocardiography is typically used to measure systolic function. Being familiar with the results of previous transthoracic echocardiograms and knowing the patient's heart rate and blood pressure during those studies is important. Anesthetic agents typically cause a decrease in preload secondary to vasodilatation, thus reducing cardiac output. With escalating doses of anesthetic medications, a reduction in contractility may be observed. Decreased cardiac output may also lead to decreased contractility because of poor coronary perfusion. Arrhythmias may develop from myocardial ischemia, compromising filling time for the LV, and thus leading to further reductions in cardiac output. A stable sinus rhythm is essential for the maintenance of adequate cardiac output.

Typical intraoperative monitoring for this patient would include an NIBP cuff and heart rate monitoring via a 5-lead ECG. Knowing the preoperative baseline vital signs is important in this patient population. During the perioperative period, including recovery, it is imperative to keep the patient near their baseline parameters. Tachycardia in particular is not well tolerated, as it decreases the filling and ejection time of the LV, thus causing decreased cardiac output. Slight hypertension is better tolerated than hypotension, as hypotension can lead to decreased coronary perfusion in an already stressed myocardium and a subsequent reduction in cardiac function.

Clinical Pearl

Tachycardia in particular is not well tolerated, as it decreases the filling and ejection time of the LV, thus causing decreased cardiac output. Slight hypertension is better tolerated than hypotension.

Is there a preferred method for postoperative pain control?

Intraoperative and postoperative pain control is of paramount importance. Inadequate pain control causes sympathetic stimulation that can increase heart rate, leading to decreased cardiac output and coronary ischemia in a patient with LV outflow tract obstruction. The specific surgical approach as well as individual patient factors will help determine which pain modalities are optimal. A multimodal approach to pain management may be achieved with a combination of preoperative oral medications, IV medications, and regional or local anesthesia. Intraoperative pain control for an umbilical hernia may be achieved with inhalational agents, IV agents such as opioids and nonsteroidal medications (ketorolac), and preoperative oral or IV acetaminophen. Regional anesthesia can also be utilized and modalities such as infiltration of local anesthesia at the surgical site by the surgeons, rectus sheath blocks, or caudal anesthesia have all been shown to be equally efficacious [10]. With umbilical hernia repair, one should aim to reduce postoperative nausea and/or retching. A reduction in the amount of opioid or an opioid-free approach may aid in decreasing this adverse event.

Can a patient with Shone complex be discharged home following hernia repair?

The appropriate postoperative disposition of congenital cardiac patients undergoing noncardiac surgery is institution, provider, and patient dependent. An umbilical hernia repair can be scheduled and safely performed as an outpatient procedure in most instances. However, in this scenario this surgery would rarely be performed at a freestanding ambulatory surgical center due to the patient's comorbidities. A preoperative discussion with the family to ensure that they are comfortable managing postoperative care and maintaining adequate pain control and hydration

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is important. This asymptomatic patient with mild to moderate obstruction, once adequately recovered from anesthesia, tolerating oral intake without difficulties and with adequate pain control, may be discharged home with parents or guardians. However, if there is concern for decreased cardiac output, hypoperfusion, or respiratory insufficiency postoperatively, the anesthesiologist and cardiologist should discuss the symptoms and an overnight stay may be warranted.

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