26 Multilevel Left-Heart Hypoplasia

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Multilevel left-heart hypoplasia (LHH) is a heterogeneous entity that involves variable underdevelopment of the mitral valve, LV, aortic valve, ascending aorta, and aortic arch. It exists within a wide spectrum that spans from hypoplastic left heart syndrome (HLHS) to mild hypoplasia of left-sided structures. The optimal management strategy for patients with a "borderline left heart" in the middle of the spectrum (i.e., single ventricle palliation [SVP] vs. biventricular repair [BVR]) is difficult to decide.

Pathophysiology and Clinical Presentation

This group of lesions, if not diagnosed prenatally, typically present in the first week of life as the PDA closes. They may present with shock (poor perfusion, tachycardia, weak pulses). Alternatively, they may be diagnosed earlier with more subtle findings such as a murmur when there is AS or a VSD, low postductal oxygen saturations on newborn pulse oximetry screening, a cardiac gallop in cases of AS, or differential pulses or BP gradient in the case of aortic coarctation. While LHH is often described as a single disease on a continuum, most lesions present as one of 3 distinct patterns, and each of these patterns likely has a different etiology of left heart hypoplasia.

Pattern 1: HLHS with Absent LV Cavity

In contrast with other forms of LHH that present with borderline left-heart structures, this pattern only presents as HLHS. In this pattern, the LV is thought to never have formed. This is the most common mechanism of mitral atresia/aortic atresia HLHS and is more thoroughly discussed in Chapter 27.

- No visible LV cavity, or only a slit-like 1-3 mm LV
- No endocardial fibroelastosis (EFE)
- Mitral atresia and aortic atresia
- Severe hypoplasia of the ascending aorta and proximal arch with more generous distal arch measurements from ductal flow filling the aortic arch retrograde throughout gestation

Pattern 2: History of Fetal AS

After a relatively normal development of the heart in utero, AS and EFE rapidly develop and the LV becomes dilated and dysfunctional. The LV and ascending aorta undergo growth arrest and are ultimately hypoplastic at birth. This postnatal presentation is highly variable, depending when the AS developed. In its most severe form, it results in HLHS (most often mitral stenosis/aortic atresia). The etiology of this pattern is unknown but is postulated to be either a primary aortic valve or myocardial disorder.

- Globular, non-apex-forming, and severely dysfunctional LV
- EFE
- Mitral stenosis with abnormal mitral valve architecture or mitral atresia
- Severe aortic valve stenosis or atresia
- The arch is typically hypoplastic

Pattern 3: The Long Skinny LV with Arch Hypoplasia/Coarctation

This pattern is sometimes called the "long, skinny pattern" or "hypoplastic left-heart complex" (HLHC). This is also sometimes referred as a "Shone-complex variant", although this a misnomer, as Shone complex describes a unique subset of these cases with supra-valvar mitral ring, parachute mitral valve, subaortic stenosis, and coarctation. The postnatal presentation is highly variable. The etiology of this pattern is unknown but it is postulated to be secondary to restricted inflow to the mitral valve and LV in utero.

- Apex-forming or near apex-forming LV that appears compressed with normal systolic function
- No EFE
- Mitral annular hypoplasia and aortic annular hypoplasia with no discrete valvar stenosis and laminar flow
- Moderate-to-severe transverse aortic arch hypoplasia with coarctation of the aorta

Approach to Management

In cases of LHH, the goal is to perform a BVR whenever possible. However, BVR is not performed when it is anticipated that patients will struggle significantly or may need a takedown back to SVP. Some patients qualify for fetal intervention to make them better BVR candidates. Postnatally, the final decision is made after echocardiographic imaging, clinical presentation, and in cases in which SVP is strongly being considered, intracardiac exploration.

Fetal Imaging and Intervention

All patients with a fetal diagnosis of LHH considered to be borderline are followed closely and counseled for a range of possible postnatal outcomes, including SVP. Some families are offered fetal intervention to make their fetus a better BVR candidate at birth. For pattern 2 (fetal AS), fetuses generally between 20 and 30 weeks gestation may be offered fetal aortic valvuloplasty if 1) the anatomic features are predicted to result in HLHS at birth and 2) aortic valvuloplasty still has the potential to stop LV-growth arrest and to improve LV function. For fetuses with pattern 3 LHH (HLHC) who are suspected to need SVP, mothers may be offered chronic maternal hyperoxygenation as part of an ongoing trial to determine if this therapy can help grow the left heart and improve neurodevelopmental outcomes.

Echocardiography

At birth, all patients are started on PGE and echocardiography is performed. Close attention is paid to LV morphology and function, mitral valve morphology, and Z-scores of all left-sided structures. When determining what treatment course to take, the type of LHH pattern (2 or 3) is seriously considered. Patients with pattern 2 LHH often have significant diastolic dysfunction and high left ventricular end-diastolic pressures that often do not improve tremendously over time. A variety of scores have been developed in the past for this pattern like the Rhodes score (Rhodes et al. 1991) to evaluate their candidacy for biventricular repair. These scores may be used in concert with clinical assessment and surgical assessment to determine treatment.

For pattern 3 LHH, most developed scores do not apply well. In fact, if they are used,

many patients would be categorized as needing SVP who would actually tolerate BVR. In pattern 3 LHH, the LV has much better postnatal growth potential, and echocardiographic measures may underestimate annular sizes due to RV compression. The large majority of these patients can successfully tolerate a BVR, even with severely hypoplastic mitral valve annuli by echocardiography. In cases in which the mitral valve or LV appear too small to undergo BVR, it may be best to try to wait at least a week after birth before intervention to reassess by echocardiography. Often, the mitral valve annulus will measure larger, the LV will be larger, and the atrial septal gradient will be lower on the second echocardiogram due to improved LV filling and improved diastolic function. This may allow for more reassurance in proceeding with a BVR. Preoperative assessment with MRI to measure flow in the ascending aorta may also be helpful to evaluate the capacity of the left-heart structures to deliver systemic cardiac output.

Surgical Considerations

One of the most important decisions that has to be made is whether the patient is suitable for a BVR or requires SVP. This complex decision involves careful analysis of all data including echocardiography and clinical status. However, echocardiographic measurements tend to underestimate the dimensions of left-heart structures in some patients due to compression by a dilated RV. As such, relying solely on echocardiographic measurements is poised to place some patients in the SVP pathway that could potentially have been adequate for BVR.

Typically, patients with LHH at TCH are not placed on a SVP pathway without an intracardiac exploration of the left heart. Intracardiac exploration includes assessment of the mitral valve size, morphology of the mitral leaflets, mitral subvalvar apparatus, size of the LV, and in some situations, size of the aortic valve. Valve size is determined based on the largest Hegar dilator that the valve easily accepts. In general, neonates with a mitral valve diameter <8 mm and an abnormal subvalvar apparatus appear to benefit from SVP. An aortic valve annulus ≥5 mm in a newborn appears to be adequate to support the systemic circulation without intervention.

In some situations, an intraoperative hemodynamic assessment is performed by temporarily occluding the PDA (thus eliminating left-to-right shunting) and assessing changes in hemodynamics, including LAP. A decrease in LAP with an increase in systemic pressure is encouraging for a BVR. In patients with a large VSD, the adequacy of the LVOT may be better assessed by placing a temporary PA band while performing a TEE.

These strategies have allowed us to offer BVR to the large majority of patients with LHH. In a recent study of 42 patients at TCH with true borderline left-heart structures, 35 (83%) patients underwent BVR (Mery et al. 2017).

The type of surgical intervention needed for patients with borderline left-heart structures is variable and depends on the specifics of each particular patient. Most patients will have some degree of involvement of the aortic arch requiring intervention to the arch early in life. The type of intervention – aortic arch reconstruction via median sternotomy (70%) vs. repair of aortic coarctation via left thoracotomy (30%) (see Chapter 25) – depends on the morphology of the aortic arch and the need to address any intracardiac anomalies at the time. Other interventions that may be required at the index operation, besides intracardiac exploration and aortic arch reconstruction,

include VSD closure, aortic valvotomy, subaortic resection, mitral valvuloplasty, aortic root replacement, or in patients with an inadequate left heart, a Norwood-type operation. Reoperations to the mitral or aortic valves later in life are common in patients undergoing BVR.

Anesthetic and Postoperative Considerations

The anesthetic management of the patient with LHH requires flexibility and good communication with the surgical team. Preoperatively, a discussion should take place regarding the need for intraoperative hemodynamic assessment and how intraoperative findings will dictate the surgical plan. The location and plan for invasive arterial pressure monitoring line placement should also be discussed; a right-radial arterial line is often preferred during aortic arch advancement to guide antegrade cerebral perfusion, and simultaneous pressure monitoring in the lower body (umbilical or femoral artery) can be helpful during coarctation repair. Extra transducers and pressure monitoring cables should be available for direct pressure monitoring.

If an intraoperative decision is made to proceed with a BVR, one must recognize that although it is a BVR, the left heart is still not normal. The LV is small and noncompliant, and diastolic dysfunction is an issue. An LAP monitoring line is often placed to guide management, especially intravascular volume management intraoperatively and postoperatively. Careful attention to the LAP can prevent volume overload and the pulmonary edema that can quickly develop in a patient with a small, noncompliant LV.

It is also essential to recognize that the valves may not be normal after CPB. In the neonatal period and in early infancy, the goal is often to repair the valve, as prosthetic valve options are severely limited. Patients may have mitral or aortic stenosis or regurgitation after CPB. Hemodynamic goals for these valvar lesions still apply.

With diastolic dysfunction and potential mitral and/or aortic stenosis, heart rate and rhythm should be monitored closely. A shortened diastole that comes with significant tachycardia limits filling, especially in a patient with a noncompliant ventricle or mitral stenosis. Likewise, dysrhythmias can be poorly tolerated, as the contribution of the atrial contraction to filling is substantial in these conditions.

Pulmonary hypertension due to preexisting LA hypertension can also be an issue perioperatively. Even after the repair of the left-sided valves, the propensity for pulmonary hypertension still exists. Care should be taken to provide a deep plane of anesthesia and to avoid hypercarbia, hypoxemia, and acidosis. Administration of milrinone or iNO should be considered to reduce PVR further.

Suggested Readings

Lara DA, Morris SA, Maskatia SA, et al. Pilot study of maternal hyperoxygenation and effect on aortic and mitral valve annular dimensions in fetuses with left heart hypoplasia. Ultrasound Obstet Gynecol 2016;48:365-372.

Mery CM, Nieto RM, De Leon LE, et al. The role of echocardiography and intracardiac exploration in the evaluation of candidacy for biventricular repair in patients with borderline left heart structures. Ann Thorac Surg 2017;103:853-861.

Moon-Grady AJ, Morris SA, Belfort M, et al. International Fetal Cardiac Intervention Registry: A Worldwide Collaborative Description and Preliminary Outcomes. J Am Coll Cardiol 2015;66(4):388-399.

Rhodes LA, Colan SD, Perry SB, et al. Predictors of survival in neonates with critical aortic stenosis. Circulation 1991;84:2325-2335.