## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Mandibular and oral soft tissue hyperplasia in acromegalics | | Airway exam<br>Check ring size | |
| CV | Hypovolemia<br>Catecholamine resistance | | Orthostatic hypotension | Give steroid replacement and observe effect on BP |
| GI | Hypoaldosteronism | Anorexia, N/V, weight loss, abdominal pain | | Hyperkalemia, hyponatremia, hypocalcemia, hypovolemia |
| ENDO | Decreased ACTH | Fatigue, fever, stress-induced hypotension, and hyponatremia | Fever, hypotension, wt loss, mental status | Morning cortisol level, rapid ACTH stimulation test, insulin tolerance test |
| | Decreased LH, FSH | Decreased libido and sexual function, amenorrhea | Regression of secondary sexual characteristics | FSH, LH serum levels, serum estradiol and testosterone |
| | Decreased GH | Fatigue | | Insulin-induced hypoglycemia, serum IGF-I |
| | Decreased TSH | Wt gain, cold intolerance, depression, constipation, hair loss | Myxedema, hyporeflexia | TSH, $T_4$ |
| | Increased prolactin | Lactation, amenorrhea | Galactorrhea | Serum prolactin |
| MS | Increased GH in acromegalics | | Large hands, feet, mandible, tongue | |
| RENAL | Increased vasopressin<br>Decreased vasopressin | Excessive thirst<br>Increased UO and thirst | <br>Hypovolemia<br>Hypotension | Hyponatremia<br>Hypernatremia<br>Dilute urine |

**Key References:** Nemergut EC, Dumont AS, Barry UT, et al.: Perioperative management of patients undergoing transsphenoidal pituitary surgery, *Anesth Analg.* 2005;101(4):1170–1181; Bajwa SJ, Kaur G: Endocrinopathies: the current and changing perspectives in anesthesia practice, *Indian J Endocr Metab.* 2015;19(4):462–469.

## Perioperative Implications

### Preoperative Preparation

- Ensure adequacy of hormone replacement therapy.
- Check serum $Na^+$, $Ca^{2+}$, and $K^+$ and correct if necessary.
- Determine volume status and adequacy of fluid replacement.
- In acromegalics, careful airway assessment and cardiac workup for possible cardiomyopathy.
- Consider steroid supplementation (hydrocortisone 100 mg/70 kg tid).
- Clinically assess for signs of increased ICP (N/V, papilledema, headache, blurry vision).

### Monitoring

- Consider arterial line if severe CV compromise, central venous pressures if indicated by inadequate preop correction of fluid status
- Monitor lytes frequently if hyponatremia or hypernatremia is not corrected preop.
- Consider glucose monitoring.

### Airway

- Acromegalic pts with normal airway exams may be difficult to intubate. Have LMA, fiberoptic, or glidescope available.
- Difficult airways in acromegalic pts due to macroglossia, hypertrophy of soft tissues of oropharynx, enlargement of soft palate, epiglottis, and aryepiglottic fold.

### Induction

- Little risk of increased ICP with pituitary adenomas.
- No special technique if hormone replacement and volume status are adequate.

### Maintenance

- Maintain normocarbia for pituitary surgery.
- Titrate narcotics and benzodiazepines carefully in pts with OSA secondary to GH-secreting tumors.

### Extubation

- Routine (for nonpituitary surgery). May need CPAP postop if pt requires use at home for possible OSA.

### Adjuvants

- Intraop DI treated with vasopressin 5 to 10 IU SQ or IM every 4–6 h

### Postoperative Period

- Polyuria and polydipsia with dilute urine may indicate development of DI.
- Postop hypopituitarism may require steroid replacement therapy.

### Anticipated Problems/Concerns

- Acromegalic pts should be treated as having difficult airways.
- Pts with GH deficiency may manifest hypoglycemia.
- Electrolyte abnormalities ($K^+$, $Na^+$, $Ca^{2+}$) and possible hypovolemia, predisposing to arrhythmias and CV instability.

# Hypoplastic Left Heart Syndrome

Gianluca Bertolizio

## Risk

- Frequency: 0.163–0.184 per 1000 live births
- 7–9% of all congenital heart diseases
- Approximately 1000 infants/y in USA
- Male to female: Up to 2:1
- 15% is associated with genetic syndromes, such as Turner syndrome, Jacobsen syndrome, trisomy 13, trisomy 18, and Smith-Lemli-Opitz syndrome

## Perioperative Risks

- 60% prenatally diagnosed (18–22 wk)
- 90% mortality within the first month without operation
- 70% overall survival to adulthood
- 30% mortality within 30 d after stage I
- 10–15% interstage mortality

## Worry About

- Premature closure of the DA and the presence of a restrictive PFO/ASD will cause a rapid decompensation of pt after birth.
- Severe hypoxemia due poor intracardial mixing or increased PVRs.
- Pulmonary edema due to pulmonary overcirculation and high LA pressure.
- Systemic and coronary hypoperfusion due to runoff (mainly diastolic) to the pulmonary circulation.
- High risk of myocardial ischemia.
- Aortic atresia + mitral atresia/stenosis subtypes have higher mortality.
- Possible presence of left ventriculocoronary fistulae.

## Overview

- Secondary to a severe hypoplasia of the left heart structures (MV, AV, LA, LV, ascending aorta, aortic arch), which leads to different degrees of obstruction of LA and/or LV outflow tract. It is defined as single ventricle physiology, which has the following characteristics:
  - Complete mixing of the Qp and Qs.
  - Ventricular output = Qp + Qs.
  - Existence of two parallel circulations: The distribution of systemic and pulmonary blood flow is dependent on the relative resistances to flow.
  - $SaO_2 = SpaO_2$.
- The goal is to manage the PVR/ SVR ratio to maintain the Qp:Qs~1.
- Qp:Qs is calculated as the following = $(SaO_2 - SvO_2) / (SpvO_2 - SpaO_2)$.
- $SvO_2$ = mix venous saturation and $SpvO_2$ = pulmonary venous saturation. $SpvO_2$ = 100% if there is no lung disease.

## Etiology

- It is a ductal-dependent circulation with complete mixing of blood through PFO or ASD.
- In a normal fetus systemic circulation, 90% of circulating blood flow is guaranteed by FO and DA. After birth, both DA and FO close. In the neonate with HLHS, blood cannot flow into the LV (MV atresia) or into the aorta (AV/aortic atresia); therefore it crosses the atrial septum through the PFO/ASD,

mixes with blood in the RA, and goes into the RV. From the RV it is pumped into the lungs via the PA and to the body via the open DA. Hence, the RV provides the cardiac output for both systemic and pulmonary circulations.

## Usual Treatment

- At birth, children are kept on $PGE_1$ to maintain the DA open and permit systemic perfusion. If FO/ASD is restrictive, it may be emergently enlarged (atrial septostomy/Rashkind).
- Functional univentricular surgical repair consists of three stages:
  - Stage I: Norwood or DKS procedure within the neonatal period—aortic arch reconstruction + systemic-to-pulmonary shunt (modified Blalock-Taussing shunt, Sano conduit are the most common). Aortic reconstruction requires DHCA/RLFP.
  - Stage II: Bidirectional Glenn procedure at 6–8 mo of age.
  - Stage III: Fontan procedure between 18 mo–3 y.
- NB: Aortic arch reconstruction may be delayed until stage II. If so stage I usually consists in a hybrid procedure = PA banding + ductal stent. Other surgical variations exist.

## Assessment Points

| System | Effect | Assessment by Hx | PE | Test |
|---|---|---|---|---|
| HEENT | Difficult intubation due to associated syndromes | | Facial dimorphism | Genetic test |
| CV | Decreased BP due to $PGE_1$<br>Poor RV function<br>Poor atrial left to right shunt due restrictive PFO/ASD<br>CHF (high LA pressure)<br>Pulm Htn due to pulm overcirculation, neonatal high PVR | Difficulty feeding<br>Need of inotropic support | Weak pulses at the lower limbs<br>Tachycardia<br>Diaphoresis<br>Hypotension<br>Tachypnea<br>Hepatomegaly<br>Metabolic acidosis | Cardiac ECHO, ECG, CXR, cardiac cath<br>Cardiac MRI |
| RESP | Apnea due to $PGE_1$<br>Normal saturation 80–85%<br>Pulm edema<br>(Qp:Qs>1)<br>Desaturation (Qp:Qs<1) | Apnea<br>Increased $O_2$ requirement<br>CPAP<br>Intubation | Respiratory failure, rales<br>Central cyanosis versus peripheral cyanosis | ABG, CXR<br>Saturation<br>$SvO_2$ |
| GI | If decreased BP: Metabolic acidosis, mesenteric ischemia, hypoglycemia NEC<br>Hypocalcemia if DiGeorge syndrome<br>Liver immaturity | Abdominal distention<br>Poor feeding<br>Blood in stool<br>Free air in peritoneum | Distended tense abdomen<br>Edema of abdominal wall<br>Tender abdomen | Coag, abdominal x-ray<br>Flank NIRS<br>LFT |
| RENAL | Renal failure secondary to decreased BP, diuretic therapy | | Oliguria | Lytes, BUN, Cr<br>UO, UA |
| CNS | Hypoglycemia, CNS hemorrhage | Increased fontanel pressure<br>Decreased Hct | Increased fontanel size and tension | Head US<br>Head NIRS |

**Key References:** Graham EM, Bradley SM, Atz AM: Preoperative management of hypoplastic left heart syndrome, *Expert Opin Pharmacother* 6(5):687–693, 2005; DiNardo JA, Zvara DA: Congenital heart disease. In DiNardo JA, Zvara DA, editors: *Anesthesia for cardiac surgery,* ed 3, Oxford, 2007, Blackwell Publishing Ltd, pp 167–251.

## Perioperative Implications

### Preoperative Preparation

- Maintain PDA patency with $PGE_1$. Atrial septostomy (ultrasound guided or in interventional radiology) may be considered to improve intracardiac mixing and decompress LA.
- Inotropic support may be needed to maintain adequate CO. Systemic perfusion can be promoted by increasing PVR (hypoxia/normoxia, mild hypercarbia, mild acidosis, increased transpulmonary pressure).
- Pt may be sedated and intubated to decrease oxygen consumption and manage the PVRs. Atelectasis should be avoided because they promote intrapulmonary shunting and V/Q mismatch.
- Aiming for $PaO_2$ = 40–45 mm Hg, saturation = 80–85%, Sa-v difference 25–30%.
- Careful evaluation of signs of systemic hypoperfusion is warranted (myocardial ischemia, splanchnic hypoperfusion, cerebral hemorrhage).

### Monitoring

- Five-lead cardiac monitoring.
- Preductal and postductal saturations.
- Cerebral NIRS ± flank NIRS.
- CVL: Know specific anatomy, including SVC variation.
- Arterial cath Usually femoral arterial line is preferred since a right mBTS will affect the right radial trace. Noninvasive BP on the upper limbs.
- Urinary cath.
- TEE.

### Airway

- Associated congenital syndromes with airway anomalies.
- Pt may be already intubated. Nasal intubation may be preferred due to better stability and if TEE placement is planned.

### Induction

- Absolute air bubble precaution.
- Induction may be carefully managed. Pt is a high risk of VT. Low aortic diastolic pressure (<30 mm Hg) and high heart rate make the heart susceptible of myocardial ischemia.
- Preinduction volume expansion may be indicated.
- Maintain HR, BP, and contractility.
- IV induction with high-dose opioids and NMB are recommended.
- Inhalational induction may be poorly tolerated and must be used judicially.
- Prolonged preoxygenation with $FIO_2$ 1.0 may cause significant increase of pulmonary flow at expense of systemic blood flow (high Qp:Qs), causing systemic hypotension and coronary hypoperfusion.

### Maintenance

- Maintain HR and contractility infusion of inotropes, such as dopamine and/or epinephrine.
- Ventilation should be adjusted to optimize PVR/SVR ratio. Usually $PaO_2$ = 40–45 mm Hg promotes minimal pulmonary vasoconstriction and provides cerebral dilatation. Low $FiO_2$ is recommended to avoid pulmonary vasodilation. Low mean airway pressures (high PIP, low RR) will decrease the transpulmonary pressure and may be needed to promote pulmonary blood flow.
- NIRS is indicated to monitor cerebral oxygenation.
- Flank NIRS may be used to monitor peripheral perfusion during DHCA/RLFP.
- A high Hct may help the splanchnic and cerebral oxygen delivery but will increase blood viscosity and PVR.
- Use of antifibrinolytic therapy should be weighed against the risk of shunt thrombosis.

### Extubation

- Deferred in PICU

### Postoperative Period

- PICU/CICU admission.
- Risk of bleeding.
- Risk of kinking/occlusion of the shunt.
- Risk of aortic obstruction.
- Maintain balanced circulation and treat low cardiac output syndrome:
  - $SaO_2$ = 75–80%, Sa-$vO_2$ = 25–30%, BP >60/30 = balanced Qp:Qs.
  - $SaO_2$ >85%, Sa-$vO_2$ >30%, decreased BP = overcirculated flow.
  - $SaO_2$ <75%, Sa-$vO_2$ 25%–30%; increased BP = undercirculated flow.
  - $SaO_2$ <70%, Sa-$vO_2$ >30%, decreased BP = low cardiac output.
- Inotropic support (to maintain CO) and milrinone (to decrease PVR and improve systemic perfusion) are often needed.
- Inhaled nitric oxide (selective pulmonary vasodilator) may be necessary.
- Diastolic pressure is usually higher after the Sano conduit placement versus other shunts.
- After Stage I Norwood repair
  - Up to 45% develop laryngeal dysfunction, dysphagia, or GERD.
  - 10%–20% NEC.
  - Risk of periop cardiac arrest for non-cardiac surgeries: 22%–27%.