Adult Congenital Heart Disease (ACHD)

What You Need to Know

- Congenital heart disease (CHD) is the most common birth defect, occurring in ~1% of births. With improved survival rates, there are now over 1.5 million adults in the US living with repaired or unrepaired CHD.
- ACHD patients are at risk for cardiovascular and systemic complications, such as: cardiomyopathy (HFpEF and HFrEF), right heart failure, pulmonary arterial hypertension, valvular heart disease, atrial and ventricular arrhythmias, and chronic lung or kidney disease.
- The 2018 AHA/ACC Guideline for Management of ACHD classifies congenital heart disease by both anatomy and physiology (Appendix A).
- Patients with severe anatomic forms of CHD, reduced functional status, and significant co-morbidities are at greater risk for complications.
- For COVID-19 Considerations in patients with ACHD, see Patients with COVID-19 and Appendix B.

Considerations by Specific ACHD Lesions

	Single Ventricle – Fontan Repair	Systemic Right Ventricle – D-transposition of the great arteries (D-TGA) s/p Mustard or Senning Repair; L-TGA	Cyanotic Congenital Heart Disease - Unrepaired or Palliated CHD
Anatomic Differences	▪ One ventricle pumping to systemic circulation (may be single right or left ventricle) ▪ SVC connects directly to right pulmonary artery (RPA) ▪ IVC is baffled through/near right atrium to RPA ▪ No right sided sub-pulmonic ventricle – all blood return to lungs is *passive*	▪ Morphologic right ventricle pumps blood to systemic circulation ▪ In Mustard/Senning repair, blood is redirected by surgical baffles	▪ Hypoxia is related to intracardiac or intrapulmonary shunting (right to left)
Oxygenation	▪ Hypoxia (baseline SpO2 < 90%) due to underlying shunts (intracardiac/ intrapulmonary) is common ▪ May be caused by: baffle fenestration, venous collaterals, pulmonary AVMs ▪ Patients usually know their baseline SpO_2 ▪ May not respond as readily to supplemental O_2 ▪ Increased risk of paradoxical embolus ▪ May have higher baseline hemoglobin	▪ In Mustard/Senning repair small shunts from baffle leaks are common, but baseline SpO_2 rarely < 90%	▪ **May not respond to supplemental oxygen** as expected ▪ Supplemental oxygen may decrease respiratory drive; goal SpO_2 should be close to baseline
Hemodynamic	▪ Patients are significantly PRELOAD dependent (have no subpulmonic ventricle) ○ **Fluid resuscitation first** with systemic hypotension or signs of low cardiac output ▪ With any drop in SVR (sedation, intubation, and sepsis) may increase right to left shunt and worsen hypoxia. **Knowing the shunt status of a patient is crucial** ▪ Document baseline O_2 saturation ▪ Less likely to respond to inotropes	▪ Systemic RV more prone to systemic dysfunction and heart failure ▪ Higher risk of heart failure if significant systemic AV valve regurgitation (tricuspid regurgitation) ▪ Systemic RV more susceptible to ischemia ▪ **May respond to inotropes**	▪ Erythrocytosis is expected ▪ May require special tubes for hematology labs (special sodium citrate – contact lab if significantly elevated hematocrit) ▪ Increased risk for bleeding diathesis AND venous thrombosis ▪ Anticoagulation NOT *routinely* recommended
Mechanical Ventilation	▪ Because all blood return to pulmonary arteries is passive (no subpulmonic ventricle), patients will have **significantly decreased cardiac output with increasing PEEP**		▪ With any drop in SVR (sedation, intubation, sepsis) may increase right to left shunt and worsen hypoxia ○ Knowing the shunt status of a patient is crucial – as hypoxia in the case of diminished SVR is likely from shunt and NOT from pulmonary disease ▪ Document baseline O_2 saturation
Invasive Monitoring	▪ Fontan anatomy may create **complexity for CVC or PA catheter placement (and possibly A-line placement)**	▪ In Mustard/Senning repair, placement of PA catheter will require fluoroscopic guidance and **should only be performed by ACHD cardiologist**	▪ Increased risk of paradoxical embolism from clots forming on CVC or PA catheter
Other	▪ **0.2 micron filters should be placed on all IVs** ▪ High risk for venous thrombus with central line	▪ Patients with baffle leaks are at increased risk for paradoxical embolism ▪ **0.2 micron filters should be placed on all IVs**	▪ Risk for paradoxical embolism (thrombus or air) ▪ **0.2 micron filters should be placed on all IVs**

Considerations by Specific ACHD Lesions			
	ACHD Pulmonary Arterial Hypertension (PAH)	**Right Heart Failure – Ebstein Anomaly, Tetralogy of Fallot, Double Outlet Right Ventricle**	**Prior Surgical Systemic to Pulmonary Artery Shunts**
Anatomic Differences	▪ PAH caused by chronic unrepaired or partially repaired intracardiac shunts (ASD, VSD, PDA) ▪ May result in PAH physiology or Eisenmenger syndrome ▪ *Eisenmenger syndrome* - Large unrepaired intracardiac shunt leading to pulmonary hypertension and chronic hypoxia/cyanosis from right to left shunt	▪ Right heart failure may occur because of intrinsic right ventricular abnormalities (Ebstein anomaly) or because of longstanding right-sided valve regurgitation	▪ Classic Blalock Taussig Thomas shunt (BTT or BT shunt) – subclavian artery is sacrificed to create communication with ipsilateral pulmonary artery
Oxygenation	▪ Chronic hypoxia is common – important to understand patient's baseline SpO_2 ▪ Can see worsening oxygenation with worsening pulmonary hypertension or decrease in systemic vascular resistance (SVR) with decreased SVR (e.g. sepsis, peripheral vasodilation) due to increased right to left shunt	▪ Should have normal oxygenation at baseline	▪ O2 saturations are dependent on intracardiac anatomy
Hemodynamic	▪ Often preload dependent, especially if severe RV dysfunction ▪ Goal systolic BP > 90 mmHg, consider early invasive arterial line ▪ For hypotension and dehydration: o *PAH physiology* – **fluid resuscitation to goal CVP 12-15,** consider: ▪ Inotropic support if severe RV dysfunction ▪ Inhaled pulmonary vasodilators (may require transfer to a unit capable of administering/monitoring these medications) o *Eisenmenger physiology* – consider pressor support, especially if worsening hypoxia	▪ Patients will be preload dependent ▪ Pre-load requirements individualized	▪ No right or left arm BP (thoracotomy scar may identify sidedness) ▪ Cuff or arterial line BPs may be inaccurate
Mechanical Ventilation	▪ Increased risk of hemodynamic instability with prone ventilation	▪ Should be the same as patients with normal cardiac anatomy	
Invasive Monitoring	▪ Increased risk of paradoxical embolism from clots forming on CVC or PA catheter ▪ Central venous line placement should be discussed with ACHD specialist	▪ If unsure of preload status, placement of central venous line for CVP monitoring is recommended	▪ Arterial line placement should be completed with ACHD assistance
Other	▪ Risk for paradoxical embolism (thrombus or air) ▪ **0.2 micron filters should be placed on all IVs** ▪ **DO NOT STOP home PAH medication without consulting ACHD/PAH specialist** o On admission, home therapies should be continued while awaiting PAH consult. If clinical need for interruption or modification is identified, contact PAH team for recommendation (Refer to Pulmonary Arterial Hypertension guideline)		

Shock Management in ACHD and PAH Conservative Therapy

- Consider vasopressor therapy with a minimal goal systolic BP > 90 mmHg (gauge SBP based on last known SPAP to maintain SBP/SPAP > 1)
- In patient with ACHD-PAH (as opposed to Eisenmenger syndrome) consider PA catheter to monitor hemodynamics and adjust/guide inotropic and/or pressor therapy
- CVP goal central pressure of ~12-14 mmHg; however, evaluating this in the context of the individual patient and weighing with pulmonary/ventilatory needs
- Follow urinary output, mental status changes and lactate to access adequacy of cardiac output and oxygenation
- Consider monitoring central venous oxygen saturation (**if no h/o shunt**), if unable to place PA catheter for estimation of Fick cardiac output to assist in the assessment and management as it relates to consideration of inotropic support
- Central venous catheter and calculation of stroke volume is an equivalent and less invasive approach to monitoring C.O. and CVP to estimate the degree and type of shock present in the hemodynamically stable patient
- ECMO evaluation is reasonable at this time (see below)

ECMO Recommendations

ECMO Cannulation

Potential Venous abnormalities:

- IVC interruption with azygous continuation
- Central venous obstruction from prior procedures
- Surgical repairs (e.g. Glenn procedure) connects SVC to RPA

Timing of ECMO

- **EARLY in the admission ACHD CT Surgery Consult** with potential for rapid deterioration
    - ACHD cardiology consult should be placed upon admission
    - Consider early consult for ACHD CT surgery for patients with ACHD to determine if there are no specific anatomic or physiologic abnormalities that warrant further investigation in the case ECMO will be considered
        - All patients with ACHD AP Classification Anatomic Stage III (Severe Complexity) and majority of II (Moderate Complexity), Appendix A, will require collaboration with ACHD CT surgery, and CT surgery
- Because of the vast complexities of patients with ACHD, timing and candidacy of ECMO includes consultation and collaboration with ACHD CT surgery service, inpatient consult to ECMO (cardiac surgery), Critical Care Anesthesiology, and ACHD cardiologist
- Standard ECMO protocol (ECMO) for indications in most circumstances when the patient meets refractory respiratory failure guidelines and/or develops vasopressor/ inotrope refractory hypotension

Airway Management – Intubation

- Only clinicians experienced with intubation should perform procedure
- Rapid sequence intubation (RSI) with paralytic - *Avoid any afterload reducing sedation as this will increase right to left shunting in the patient with a residual defect and/or fenestration*
- Video laryngoscopy may allow operator more distance from airway
- Bougie may increase first pass success

Shock Management

- Vasopressor support to keep MAP > 60-65 mmHg:
    - **1st line agent**: Vasopressin (particularly in patients with PAH or Fontan)
    - **2nd line agent**: Norepinephrine
    - **3rd line agent**: Epinephrine

  NOTE: If any of these agents are not available in the Pyxis, place an order and contact pharmacy to have them delivered to the bedside at the time of intubation, in case hypotension develops during induction
- In severe RV dysfunction, consider pre-intubation CVL placement, volume resuscitation with goal CVP of 10-15 mmHg

Patients with COVID-19

Consider limited echo for LVEF (to evaluate for myocarditis) if it will change management

ARDS should be facilitated by pulmonary medicine consult and includes the following:

- Support positive end expiratory pressure (PEEP) to maintain alveolar patency with low tidal volume
    - **CAUTION**: Patients with **single ventricle and Fontan palliation** are highly pre-load dependent with no subpulmonic ventricle and therefore will demonstrate significantly reduced cardiac output with increasing PEEP
    - It is likely that a patient with this anatomy should be targeted to the lower PEEP range when increased positive end expiratory pressure is required for oxygenation

For additional information, refer to the Airway Management guideline (link pending)

References

- Ginde, Salil, et al. (2013) "Restrictive lung disease is an independent predictor of exercise intolerance in the adult with congenital heart diease." *Congenital heart disease*. 8.3: 246-254.
- Kuijpers, Joey, M., et al. (2020) "Risk of coronary artery disease in adults with congenital heart disease: A comparison with the general population." *International journal of cardiology*. 304:39-42.
- Stout, Karen K. and Daniels CJ et al. (2019) "2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines." *Journal of the American College of Cardiology*. 73.12: 6370697.

OSUWMC Resources

- IHIS Admission Notification for Providers of ACHD pts.

Quality Measures

- Number of patients admitted by lesion type

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Disclaimer: *Clinical practice guidelines and algorithms at The Ohio State University Wexner Medical Center (OSUWMC) are standards that are intended to provide general guidance to clinicians. Patient choice and clinician judgment must remain central to the selection of diagnostic tests and therapy. OSUWMC's guidelines and algorithms are reviewed periodically for consistency with new evidence; however, new developments may not be represented.*

Appendix A: ACHD AP Classification

CHD Anatomy

I: Simple

Native disease

- Isolated small ASD
- Isolated small VSD
- Mild isolated pulmonic stenosis

Repaired conditions

- Previously ligated or occluded ductus arteriosus
- Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement

II: Moderate Complexity

Repaired or unrepaired conditions

- Aorto-left ventricular fistula
- Anomalous pulmonary venous connection, partial or total
- Anomalous coronary artery arising from the pulmonary artery
- Anomalous aortic origin of a coronary artery from the opposite sinus
- AVSD (partial or complete, including primum ASD)
- Congenital aortic valve disease
- Congenital mitral valve disease Coarctation of the aorta
- Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
- Infundibular right ventricular outflow obstruction
- Ostium primum ASD
- Moderate and large unrepaired secundum ASD
- Moderate and large persistently patent ductus arteriosus
- Pulmonary valve regurgitation (moderate or greater)
- Pulmonary valve stenosis (moderate or greater)
- Peripheral pulmonary stenosis
- Sinus of Valsalva fistula/aneurysm
- Sinus venosus defect
- Subvalvar aortic stenosis (excluding HCM)
- Supravalvar aortic stenosis
- Straddling atrioventricular valve
- Repaired tetralogy of Fallot
- VSD with associated abnormality and/or moderate or greater shunt

III. Great Complexity (or Complex)

- Cyanotic congenital heart defect (unrepaired or palliated, all forms)
- Double-outlet ventricle
- Fontan procedure
- Interrupted aortic arch
- Mitral atresia
- Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)
- Pulmonary atresia (all forms)
- TGA (classic or d-TGA; CCTGA or I-TGA)
- Truncus arteriosus
- Other abnormalities of atrioventricular and ventriculoarterial connection (i.e. crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Physiological Stage

A

- NYHA FC I symptoms
- No hemodynamic or anatomic sequelae
- No arrhythmias
- Normal exercise capacity
- Normal renal/hepatic/pulmonary function

B

- NYHA FC II symptoms
- Mild hemodynamic sequela (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction)
- Mild valvular disease
- Trivial or small shunt (not hemodynamically significant)
- Arrhythmia not requiring treatment
- Abnormal objective cardiac limitation to exercise

C

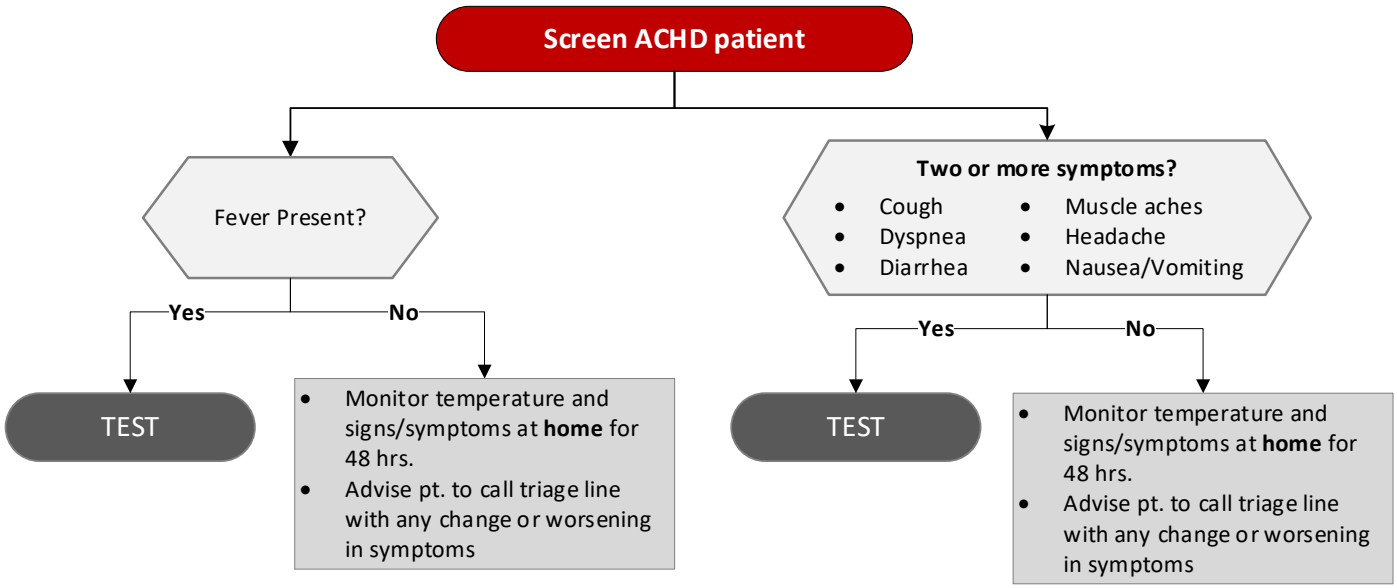
- NYHA FC III symptoms
- Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonic, or both)
- Moderate aortic enlargement
- Venous or arterial stenosis
- Mild or moderate hypoxemia/cyanosis
- Hemodynamically significant shunt
- Arrhythmias controlled with treatment
- Pulmonary hypertension (less than severe)
- End-organ dysfunction responsive to therapy

D

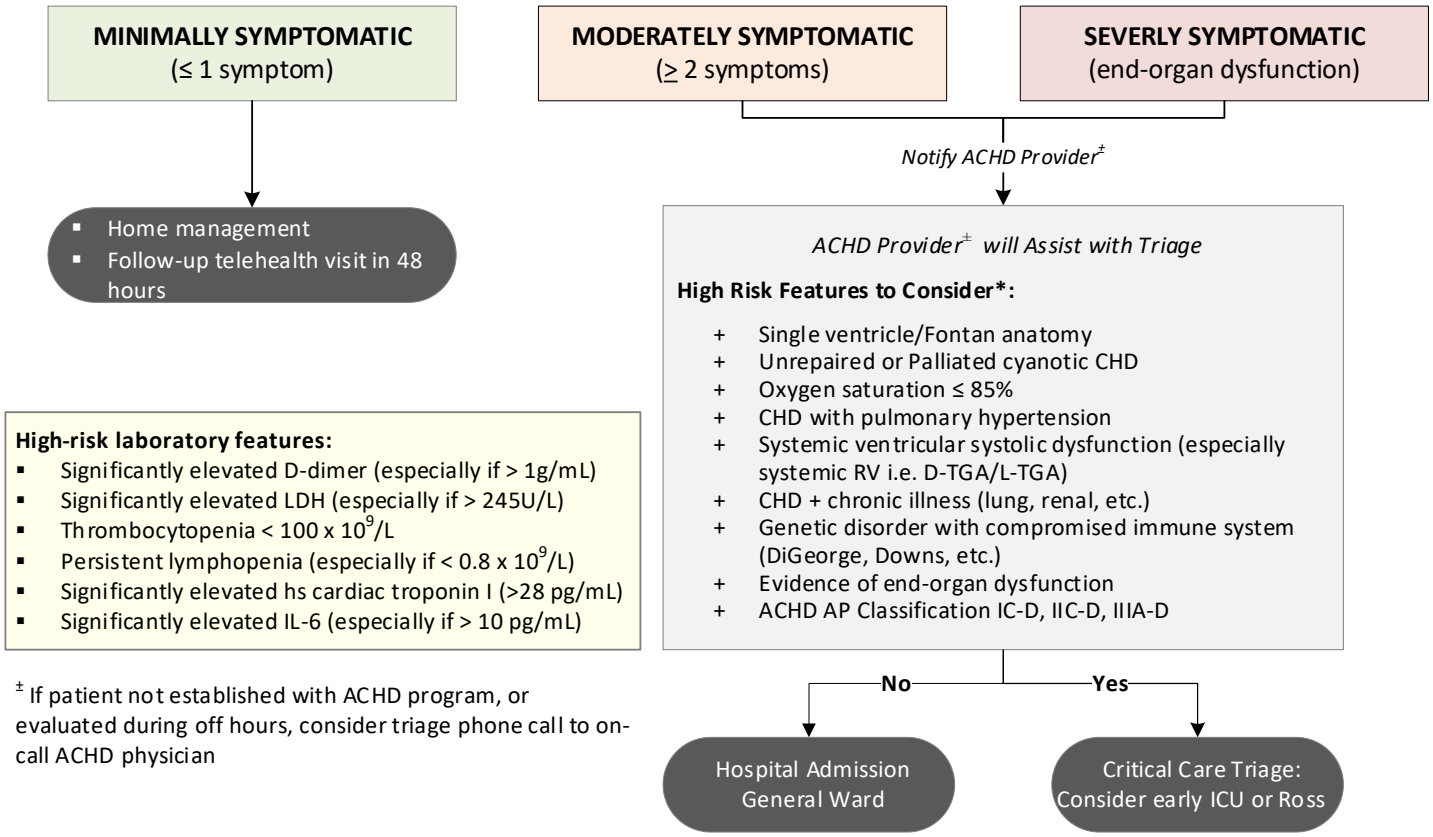
- NYHA FC IV symptoms
- Severe aortic enlargement
- Arrhythmias refractory to treatment
- Severe hypoxemia (almost always associated with cyanosis)
- Severe pulmonary hypertension
- Eisenmenger syndrome
- Refractory end-organ dysfunction

Appendix B: ACHD Patients with Suspected/Confirmed COVID-19

Algorithm 1: COVID-19 Screening



Algorithm 2: Triage for ACHD Patients with Suspected COVID-19



High-risk laboratory features:

- Significantly elevated D-dimer (especially if > 1g/mL)
- Significantly elevated LDH (especially if > 245U/L)
- Thrombocytopenia < 100 x 10^9/L
- Persistent lymphopenia (especially if < 0.8 x 10^9/L)
- Significantly elevated hs cardiac troponin I (>28 pg/mL)
- Significantly elevated IL-6 (especially if > 10 pg/mL)

± If patient not established with ACHD program, or evaluated during off hours, consider triage phone call to on-call ACHD physician