# CT Children's CLASP Guideline

## *Connecticut Adult Congenital Heart Service (CTACH)*

### INTRODUCTION

Congenital heart disease (CHD) is the most common birth defect, accounting for ~1% of all live births. 20,000-40,000 children with CHD graduate to adulthood each year. There are over 1.4 million adults with congenital heart disease living in the USA. Adults with CHD have been shown to have better outcomes when followed in a specialized adult congenital heart disease (ACHD) center. CT Children's became the first accredited ACHD program in the state in 2018.

### INITIAL EVALUATION AND MANAGEMENT

**INITIAL EVALUATION:**

- The focus of the initial evaluation by the referring provider should be on classifying the CHD into mild, moderate, or great complexity. This determines where the patient should receive care, and how often. ***(See Appendix: Referral Guidelines to CTACH)***
- Obtain routine blood work, ECG per referring provider discretion
- All other testing (echocardiogram, cardiac MRI, cardiac cath, EP evaluation, etc.) is preferred to be completed through Connecticut Adult Congenital Heart Service (CTACH)

### WHEN TO REFER

**URGENT REFERRAL to Connecticut Children's CTACH Service:** (will be seen within 1 week or less) for any of the following, regardless of complexity of CHD:

- Acute cardiac issues: Worsening of pre-existing arrhythmia or new arrhythmia, symptoms of heart failure, or worsening cyanosis
- Worsening of pre-existing pulmonary hypertension (PH) or new development of PH
- Patients who become pregnant or plan on becoming pregnant

**ROUTINE REFERRAL to Connecticut Children's CTACH Service:**
(initial consultation within 4 weeks) for:

- **Mild complexity CHD**: patient can be managed by their general cardiologist or internist. CTACH providers can provide support and advice to the referring provider by phone, or can see patients for consultation as needed
- **Moderate complexity CHD**: can be co-managed. CTACH program will manage the congenital abnormalities, while the general adult cardiologist can manage coronary disease, hypertension, hyperlipidemia, etc. Management of heart failure is done collaboratively
- **Great complexity CHD**: Cardiac care should primarily occur within the CTACH program

### HOW TO REFER

**Referral to Connecticut Adult Congenital Heart (CTACH) Disease Service via**
**CT Children's One Call Access Center**
For more information on how to place referrals to Connecticut Children's, click **here**.
**Phone:** 833.733.7669 **Fax:** 833.226.2329

***Information to be included with the referral:***

- Most recent office visit note
- Available copies of ECG, echocardiogram or other imaging data, preferably with image CDs and operative reports

### WHAT TO EXPECT

**What to expect from CT Children's Visit:**

- Outpatient clinic visit with an adult congenital specialist. We have 3 board certified ACHD doctors and 1 highly trained ACHD APRN
- In office testing, including an ECG and an echocardiogram, are completed during the first appointment
- Subsequent testing such as a cardio-pulmonary exercise testing, cardiac MRI, right heart cath, and EP study will be scheduled as needed based on the initial visit

## APPENDIX: Referral Guideline to Connecticut Adult Congenital Heart Service (CTACH)

| Mild Complexity CHD | Moderate Complexity CHD | Great Complexity CHD |
|---|---|---|
| **Native disease**<br>1. Isolated congenital aortic valve disease<br>2. Isolated congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)<br>3. Small atrial septal defect<br>Isolated small ventricular septal defect (no associated lesions)<br>4. Mild pulmonary stenosis<br>Small patent ductus arteriosus<br><br>**Repaired conditions**<br>1. Previously ligated or occluded ductus arteriosus<br>2. Repaired secundum or sinus venosus atrial septal defect without residua<br>3. Repaired ventricular septal defect without residua | 1. Aorto–left ventricular fistulas<br>2. drainage Partial or total<br>3. Atrioventricular septal defects (partial or complete)<br>4. Coarctation of the aorta<br>5. Ebstein's anomaly<br>6. Infundibular right ventricular outflow obstruction of significance<br>7. Ostium primum atrial septal defect<br>8. Patent ductus arteriosus (not closed)<br>9. Moderate to severe pulmonary valve regurgitation or stenosis<br>10. Sinus of Valsalva fistula/aneurysm<br>11. Sinus venosus atrial septal defect<br>12. Subvalvular AS or Supra AS (except HOCM)<br>13. Tetralogy of Fallot<br>14. Ventricular septal defect with:<br>- Absent valve or valves<br>- Aortic regurgitation<br>- Coarctation of the aorta<br>- Mitral disease<br>- Right ventricular outflow tract obstruction<br>- Straddling tricuspid/mitral valve<br>- Subaortic stenosis | 1. Conduits, valved or non-valved<br>2. Cyanotic congenital heart (all forms)<br>3. Double-outlet ventricle<br>4. Eisenmenger syndrome<br>5. Fontan procedure<br>6. Mitral atresia<br>7. Single ventricle (also called double inlet or outlet, common, or primitive)<br>8. Pulmonary atresia (all forms)<br>9. Pulmonary vascular obstructive disease<br>10. Transposition of the great arteries<br>11. Tricuspid atresia<br>12. Truncus arteriosus/hemitruncus<br>13. Other abnormalities of atrio-ventricular or ventriculo-arterial connection not included above:,<br>- Crisscross heart<br>- L-isomerism<br>- Heterotaxy syndromes<br>- Ventricular inversion (L-transposition) |
| LOCUS OF CARE RESTS WITH GENERAL CARDIOLOGY<br>Phone/email/clinic consultation with CTACH if needed | CO-MANAGEMENT BETWEEN GENERAL CARDIOLOGY AND CTACH<br>Annual follow-up, alternating with CTACH or sooner as needed | PRIMARY MANAGEMENT BY CTACH, CONSULTATION WITH GENERAL CARDIOLOGY AS NEEDED<br>Annual CTACH evaluation or sooner as needed |

**Please call to discuss whether referral is needed and/or if diagnoses do not fall under above classification, refer to CTACH service for determination of complexity and further management.**