




CT Children's CLASP Guideline

CT Adult Congenital Heart Service (CTACH)

INTRODUCTION	<p>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 (more than children with CHD). Adults with CHD (ACHD) can be classified into mild, moderate, and great complexity. Moderate and great complexity patients should be followed by a physician with expertise in ACHD.</p>
INITIAL EVALUATION AND MANAGEMENT	<p>INITIAL EVALUATION:</p> <ul style="list-style-type: none"> ▪ The focus of the initial evaluation should be on classifying the CHD into mild, moderate, or great complexity. <i>(See Appendix: Referral Guidelines to CTACH)</i> ▪ If appropriate, obtain routine blood work, ECG. ▪ All other testing is preferred to be completed through Connecticut Adult Congenital Heart Service (CTACH).
WHEN TO REFER	<p>URGENT REFERRAL to Connecticut Children's CTACH Service: (within 1 week or less) for any of the following, regardless of complexity of CHD:</p> <ul style="list-style-type: none"> ▪ Acute cardiac issues: Worsening of pre-existing arrhythmia or new arrhythmia, symptoms of heart failure or cyanosis ▪ Worsening of pre-existing pulmonary hypertension (PH) or new development of PH ▪ Patients who become pregnant or plan on becoming pregnant <p>ROUTINE REFERRAL to Connecticut Children's CTACH Service: (initial consultation within 4 weeks) for:</p> <ul style="list-style-type: none"> ▪ Mild complexity CHD: patient can be managed by their general cardiologist. Phone, email consultations or referral as deemed necessary by primary general cardiologist. ▪ Moderate complexity CHD: patient should be evaluated by CTACH once every 1-2 years depending on clinical status, or urgently, if needed, as above. Management of coronary artery disease, hyperlipidemia will be as per general cardiology. ▪ Great complexity CHD: patient should be evaluated by CTACH once per year, or urgently, if needed, as above.
HOW TO REFER	<p>Referral to Connecticut Adult Congenital Heart (CTACH) Disease Service via CT Children's One Call Access Center Phone: 833.733.7669 Fax: 833.226.2329</p> <p>Information to be included with the referral:</p> <ul style="list-style-type: none"> ▪ Most recent clinic letter ▪ Available copies of ECG, Echocardiogram or other imaging data preferably with image CDs and operative reports
WHAT TO EXPECT	<p>What to expect from CT Children's Visit:</p> <ul style="list-style-type: none"> ▪ Outpatient clinic visit with an adult congenital cardiologist ▪ Testing including an ECG and an echocardiogram ▪ Subsequent testing such as a cardio-pulmonary exercise testing or a cardiac MRI to be determined based on the initial visit

**Any CHD patient seen by an internist should be referred to general adult cardiology or directly to CTACH for a determination of the complexity category and decision regarding follow-up care.*

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

Mild Complexity CHD	Moderate Complexity CHD	Great Complexity CHD
<p style="text-align: center;"><u>Native disease</u></p> <ol style="list-style-type: none"> 1. Isolated congenital aortic valve disease 2. Isolated congenital mitral valve disease (eg, except parachute valve, cleft leaflet) 3. Small atrial septal defect Isolated small ventricular septal defect (no associated lesions) 4. Mild pulmonary stenosis Small patent ductus arteriosus <p style="text-align: center;"><u>Repaired conditions</u></p> <ol style="list-style-type: none"> 1. Previously ligated or occluded ductus arteriosus 2. Repaired secundum or sinus venosus atrial septal defect without residua 3. Repaired ventricular septal defect without residua <div style="text-align: center; margin-top: 20px;">  <div style="border: 2px solid green; padding: 10px; background-color: #00b050; color: white; text-align: center; width: fit-content; margin: 0 auto;"> <p>LOCUS OF CARE RESTS WITH GENERAL CARDIOLOGY</p> </div> <div style="border: 1px solid green; padding: 5px; background-color: white; text-align: center; width: fit-content; margin: 5px auto;"> <p>Phone/email/clinic consultation with CTACH if needed</p> </div> </div>	<ol style="list-style-type: none"> 1. Aorto–left ventricular fistulas drainage Partial or total 2. Atrioventricular septal defects (partial or complete) 3. Coarctation of the aorta 4. Ebstein’s anomaly 5. Infundibular right ventricular outflow obstruction of significance 6. Ostium primum atrial septal defect 7. Patent ductus arteriosus (not closed) 8. Moderate to severe pulmonary valve regurgitation or stenosis 9. Sinus of Valsalva fistula/aneurysm 10. Sinus venosus atrial septal defect 11. Subvalvular AS or Supra AS (except HOCM) 12. Tetralogy of Fallot 13. Ventricular septal defect with: <ul style="list-style-type: none"> - Absent valve or valves - Aortic regurgitation - Coarctation of the aorta - Mitral disease - Right ventricular outflow tract obstruction - Straddling tricuspid/mitral valve - Subaortic stenosis <div style="text-align: center; margin-top: 20px;">  <div style="border: 2px solid orange; padding: 10px; background-color: #ff7f0e; color: white; text-align: center; width: fit-content; margin: 0 auto;"> <p>CO-MANAGEMENT BETWEEN GENERAL CARDIOLOGY AND CTACH</p> </div> <div style="border: 1px solid orange; padding: 5px; background-color: white; text-align: center; width: fit-content; margin: 5px auto;"> <p>Annual follow-up, alternating with CTACH or sooner as needed</p> </div> </div>	<ol style="list-style-type: none"> 1. Conduits, valved or non-valved 2. Cyanotic congenital heart (all forms) 3. Double-outlet ventricle 4. Eisenmenger syndrome 5. Fontan procedure 6. Mitral atresia 7. Single ventricle (also called double inlet or outlet, common, or primitive) 8. Pulmonary atresia (all forms) 9. Pulmonary vascular obstructive disease 10. Transposition of the great arteries 11. Tricuspid atresia 12. Truncus arteriosus/hemitruncus 13. Other abnormalities of atrio-ventricular or ventriculo-arterial connection not included above; <ul style="list-style-type: none"> - Crisscross heart - L-isomerism - Heterotaxy syndromes - Ventricular inversion (L-transposition) <div style="text-align: center; margin-top: 20px;">  <div style="border: 2px solid red; padding: 10px; background-color: #d62728; color: white; text-align: center; width: fit-content; margin: 0 auto;"> <p>PRIMARY MANAGEMENT BY CTACH, CONSULTATION WITH GENERAL CARDIOLOGY AS NEEDED</p> </div> <div style="border: 1px solid red; padding: 5px; background-color: white; text-align: center; width: fit-content; margin: 5px auto;"> <p>Annual CTACH evaluation or sooner as needed</p> </div> </div>
<p>Please call to discuss whether referral is needed and/or if diagnoses do not fall under above classification, refer to CTACH service for further determination of complexity and further management.</p>		

