

Date: December 3, 2012

Title: Care of Adults with Congenital Heart Disease

Clinical Question:

- P (*population*) Among healthcare facilities, and its healthcare providers, performing cardio-thoracic surgery and other cardiovascular procedures for adults with congenital heart disease (ACHD),
- I (*intervention*) does care provided at a pediatric facility versus
- C (*comparison*) care provided at an adult facility
- O (*outcome*) lead to better patient outcomes (e.g. lower post-op mortality rate, fewer adverse events)?

[Definitions for terms marked with * may be found in the Supporting Information section.](#)

Target Population for the Recommendation:

Adults with congenital heart disease, 18 years of age and older, who require cardio-thoracic surgery, and other cardiovascular procedures specific to their congenital heart defect.

Recommendation: It is recommended that cardio-thoracic surgery and other cardiovascular procedures for ACHD patients be performed in an ACHD regional center which may be established in a pediatric facility, adult facility, combined or freestanding unit (American College of Cardiology, 2001 [5a]; Warnes, 2008 [5a]; Murphy, 2003 [5a]; Landzberg, 2001 [5a]; Deanfield, 2003 [5a]; Ochiai, 2011 [5a]; Webb, 2010 [5a]).

Discussion/Summary of Evidence Related to the Recommendation:

Several expert opinion articles addressed the PICO question by describing the optimal care paradigm in which ACHD patients should receive care. As stated in the 32nd Bethesda Conference Report (Landzberg, 2001 [5a]) and repeated throughout expert opinion articles (Deanfield, 2003 [5a]; Ochiai, 2011 [5a]; Webb, 2010 [5a]), ACHD care should be organized into a regional and national system of specialized ACHD centers similar to the Severe Heart Failure Model of Care. According to this model, care would be delivered to ACHD patients through local medical resources which would refer patients to specific regional centers for specialized care which includes cardio-thoracic surgery and other cardiovascular procedures and inpatient care. However, there are no “regional centers” located in the United States; only ACHD clinics which are housed in adult or pediatric facilities (Patel, 2010 [5a]). The American College of Cardiology/American Heart Association (ACA/AHA) recommends that specialized regional centers be created to manage ACHD that would serve as a resource and link to surrounding medical centers, patients and their families (Warnes et al., 2008 [5a]).

Dimensions for Judging the Strength of the Recommendation:

Reflecting on your answers to the dimensions below and given that more answers to the left of the scales indicates support for a stronger recommendation, complete one of the sentences above to judge the strength of this recommendation.

(Note that for negative recommendations, the left/right logic may be reversed for one or more dimensions.)

1. Grade of the Body of Evidence	<input type="checkbox"/> High	<input type="checkbox"/> Moderate	<input checked="" type="checkbox"/> Low
Comments:			
2. Safety/Harm (Side Effects and Risks)	<input type="checkbox"/> Minimal	<input checked="" type="checkbox"/> Moderate	<input type="checkbox"/> Serious
Comments: Implementation of ACHD Regional Centers similar to the Severe Heart Failure Model of care may afford patients fewer hospital visits and improved medical and surgical outcomes (Landzberg, 2001 [5a]).			
3. Health benefit to patient	<input type="checkbox"/> Significant	<input checked="" type="checkbox"/> Moderate	<input type="checkbox"/> Minimal
Comments: Treatment in ACHD regional centers will positively influence the health outcomes of ACHD patients (Landzberg, 2001 [5a]).			
4. Burden on patient to adhere to recommendation	<input type="checkbox"/> Low	<input checked="" type="checkbox"/> Unable to determine	<input type="checkbox"/> High
Comments:			
5. Cost-effectiveness to healthcare system	<input type="checkbox"/> Cost-effective	<input checked="" type="checkbox"/> Inconclusive	<input type="checkbox"/> Not cost-effective
Comments: Evidence does not directly address cost-effectiveness to care in children's hospitals as compared to adult hospitals. Because there are no regional centers located in the United States, studies to cost-effectiveness cannot be done. Therefore, no determination regarding cost-effectiveness could be made.			
6. Directness of the evidence for this target population	<input checked="" type="checkbox"/> Directly relates	<input type="checkbox"/> Some concern of directness	<input type="checkbox"/> Indirectly relates
Comments:			
7. Impact on morbidity/mortality or quality of life	<input type="checkbox"/> High	<input checked="" type="checkbox"/> Medium	<input type="checkbox"/> Low
Comments: Quality of life would be improved if ACHD Regional Centers were implemented (Landzberg, 2001 [5a]). Landzberg (2001 [5a]) also states that if ACHD Regional Centers are established similar to the Severe Heart Failure Model of care patients may experience improved medical and surgical outcomes which may mean decreased morbidity and mortality rates. More research needs to be done in order to determine morbidity/mortality rates in ACHD Regional Centers.			

Reference List: (Evidence Level in []; See Table of Evidence Levels following references)

- American College of Cardiology. 32nd Bethesda Conference: Care of the adult with congenital heart disease.(2001). *Journal of the American College of Cardiology*, 37, 1161-1198. [5a]
- Deanfield, J. J. (2003). Management of grown up congenital heart disease. *European Heart Journal*, 24(11), 1035-1084. [5a]
- Landzberg, M. J., Murphy Jr., D. J., Davidson Jr., W. R., Jarcho, J. A., Krumholz, H. M., Mayer Jr., J. E., et al. (2001). Task force 4: Organization of delivery systems for adults with congenital heart disease. *Journal of the American College of Cardiology*, 37(5), 1187-1193. [5a]
- Murphy, D. J. (2003). The patient population and requirements for optimal care. *Progress in Pediatric Cardiology*, 17(1), 3-7. [5a]
- Ochiai, R., Murakami, A., Toyoda, T., Kazuma, K., & Niwa, K. (2011). Opinions of physicians regarding problems and tasks involved in the medical care system for patients with adult congenital heart disease in japan. *Congenital Heart Disease*, 6(4), 359-365.[5a]
- Patel, M. S., & Kogon, B. E. (2010). Care of the adult congenital heart disease patient in the United States: A summary of the current system. *Pediatric Cardiology*, 31(4), 511-514. [5a]
- Warnes, C. A., Williams, RG., Bashore, T. M., Child, J. S., Connolly, H. M., Dearani, J. A., et al. (2008). ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: Executive summary: A report of the American College of Cardiology/American Heart Association task force on practice guidelines (writing committee to develop guidelines for the management of adults with congenital heart disease). *Circulation*, 118(23), 2395-2451. [5a]
- Webb, G. (2010). The long road to better ACHD care. *Congenital Heart Disease*, 5(3), 198-205. [5a]

SUPPORTING INFORMATION

Background/Purpose of BEST Development:

When ACHD present with anatomic/physiologic problems related to CHD residua the issue of where to obtain the best medical and/or surgical care becomes pertinent. At pediatric facilities congenital heart surgeons and pediatric care staff are familiar with the anatomy and medical sequelae of CHD, but may lack expertise on how to care for adults and their accompanying comorbidities. Conversely, staff at an adult facility may have knowledge of comorbidities and adult care, but lack expertise on how to best care for the intricacies of CHD. Subsequently, many CHD patients are lost to follow up care as adults because they are no longer pediatric patients and they do not know where to go to get the best care. Pediatric healthcare professionals at Cincinnati Children's Hospital Medical Center (CCHMC) currently treat ACHD patients at the Adult Heart Clinic. These patients also come to CCHMC when surgical and other cardiovascular interventions are needed.

Definitions:

Cardiovascular procedures include any medical procedure that involves evaluation of the cardiovascular system such as, cardiac catheterizations, cardiac echocardiograms, or exercise tests.

A pediatric facility is defined as a facility where specialized congenital heart surgeons (or pediatric surgeons) practice and direct-care staff are trained to care for children.

An adult facility is defined as a facility where general heart surgeons practice and direct-care staff are trained to care for adults.

Applicability Issues:

If an ACHD regional center were to be established within CCHMC several applicability issues would have to be addressed. The location of the regional center can be within the current building or a freestanding unit on the hospital campus. If the regional center is to be established in the current building a decision should be made whether to treat ACHD patients in the current Cardiac Intensive Care Unit and Cardiac Step-Down Unit or a separate unit dedicated to ACHD patients. The option of training current staff or hiring a separate group of staff to care for this population should be decided.

Because ACHD patients are a fairly new population the following should also be considered:

- Development of educational materials for staff and ACHD patients
- Creating clinical pathways and order sets
- Properly training staff
- Creating/applying policies and procedures and Standards of Care related to ACHD patient care

Applying these concepts will involve stakeholder input, capital investment and research.

Outcome or Process Measures:

Mortality rates among ACHD patients undergoing cardiac surgery and other cardiovascular procedures
 Morbidity rates among ACHD patients undergoing cardiac surgery and other cardiovascular procedures
 Adverse events among ACHD patients undergoing cardiac surgery and other cardiovascular procedures
 Readmission rates among ACHD patients undergoing cardiac surgery and other cardiovascular procedures
 Satisfaction with care received among ACHD patients
 Direct care staff satisfaction with processes in place to care for the ACHD patient

Search Strategy:

Databases Searched: Scopus

Search Terms: congenital heart defects; cardiac patients, hospital, adult and child; pediatric nursing, adult care; congenital heart disease, adult and pediatric facility; ACHD, congenital heart surgeon, adult heart surgeon

Filters: English Language

Last Search: August 17, 2012

Relevant CCHMC Evidence-Based Documents:

None were found

Group/Team Members:

Team Leader/Author- Jeanette Harris, MBA, BSN, BS, RN

Support/Consultant- Carolyn Smith, MSN/RN, Center for Professional Excellence – Research & Evidence Based Practice

Conflicts of Interest were declared for each team member:

- No financial conflicts of interest were found.
 The following financial conflicts of interest were disclosed:

Note: Full tables of evidence grading system available in separate document:

- [Table of Evidence Levels of Individual Studies by Domain, Study Design, & Quality](#) (abbreviated table below)
- [Grading a Body of Evidence to Answer a Clinical Question](#)
- [Judging the Strength of a Recommendation](#) (abbreviated table below, dimensions table above)

Table of Evidence Levels (see note above)

Quality level	Definition
1a ⁺ or 1b ⁺	Systematic review, meta-analysis, or meta-synthesis of multiple studies
2a or 2b	Best study design for domain
3a or 3b	Fair study design for domain
4a or 4b	Weak study design for domain
5a or 5b	General review, expert opinion, case report, consensus report, or guideline
5	Local Consensus

†a = good quality study; b = lesser quality study

Table of Recommendation Strength (see note above)

Strength	Definition
It is strongly recommended that... It is strongly recommended that... not...	There is consensus that benefits clearly outweigh risks and burdens (or visa-versa for negative recommendations).
It is recommended that... It is recommended that... not...	There is consensus that benefits are closely balanced with risks and burdens.
There is insufficient evidence and a lack of consensus to make a recommendation...	

Copies of this Best Evidence Statement (BEST) and related tools (if applicable, e.g., screening tools, algorithms, etc.) are available online and may be distributed by any organization for the global purpose of improving child health outcomes.

Website address: <http://www.cincinnatichildrens.org/svc/alpha/h/health-policy/best.htm>

Examples of approved uses of the BEST include the following:

- copies may be provided to anyone involved in the organization's process for developing and implementing evidence based care;
- hyperlinks to the CCHMC website may be placed on the organization's website;
- the BEST may be adopted or adapted for use within the organization, provided that CCHMC receives appropriate attribution on all written or electronic documents;
- copies may be provided to patients and the clinicians who manage their care.

Notification of CCHMC at EBDMinfo@cchmc.org for any BEST adopted, adapted, implemented, or hyperlinked by the organization is appreciated.

Please cite as: Harris, J., Cincinnati Children's Hospital Medical Center, Best Evidence Statement: Care of Adults with Congenital Heart Disease, <http://www.cincinnatichildrens.org/svc/alpha/h/health-policy/best.htm>, BEST 126, pages 1-5, 12/3/12.

This Best Evidence Statement has been reviewed against quality criteria by 2 independent reviewers from the CCHMC Evidence Collaboration.

For more information about CCHMC Best Evidence Statements and the development process, contact the Evidence Collaboration at EBDMinfo@cchmc.org.

Note

This Best Evidence Statement addresses only key points of care for the target population; it is not intended to be a comprehensive practice guideline. These recommendations result from review of literature and practices current at the time of their formulation. This Best Evidence Statement does not preclude using care modalities proven efficacious in studies published subsequent to the current revision of this document. This document is not intended to impose standards of care preventing selective variances from the recommendations to meet the specific and unique requirements of individual patients. Adherence to this Statement is voluntary. The clinician in light of the individual circumstances presented by the patient must make the ultimate judgment regarding the priority of any specific procedure.