The 32nd Bethesda Conference Guidelines—Highlights for the Patient

Task Force 1: The Changing Profile of Congenital Heart Disease in Adult Life

- There are more adults with congenital heart defects than ever before. This is due in part to new and improved surgeries. There are an estimated 750,000 adults with congenital heart disease (CHD) in the United States. More than half need on-going specialized care.

- Studies on children show that death rates from CHD surgeries are higher at centers that do not regularly do CHD operations. This is likely to apply to surgeries in adults.

- There are many diagnostic tools for CHD. These include catherization, echocardiography, CT scans, and cardiac MRI. One study found that 53% of CHD echocardiograms done at local centers had errors. ACHD patients should get these studies done at centers that specialize in CHD.

Task Force 2: Special Health Care Needs of Adults with Congenital Heart Disease

Managing the Transition to Adulthood

- Adults with congenital heart defects (ACHDers) need help switching from pediatric to adult care. A regional ACHD center that can meet patient's needs should be identified (see task force 4). This center should offer the same quality or higher quality care than the pediatric center.

- Many factors can prevent adults with CHD from continuing their cardiac care. These include lack of symptoms and the idea that their CHD is "cured". Some are attached to their existing centers and do not want to make a change.

- All ACHD patients need education on their condition. Topics should include:

    Name and details of their heart condition
    Surgeries that have been performed
    Frequency of follow-up
    Medications and side effects
    Preventing heart infections (endocarditis)
    Exercise
    Birth control and family planning
    Career planning and resources
    Insurance
    Dental care
    Symptoms to look for
    When to contact their health care provider.

- Patients should keep a copy of their health information. This should include test results and information on operations. A shorter "health passport" should be developed. This passport should be taken along when the patient travels.

- Most adults with CHD can work. Adolescents with CHD should receive appropriate career counseling. Most should be encouraged to pursue higher education. Guidance should be given on any physical limits that may interfere with job choices.

Unique Medical Issues

- Cyanotic ("blue") patients have many special health needs. Problems can occur throughout the body, not just in the heart and lungs. Primary care doctors should be made aware of the special needs of these patients. They should be seen at ACHD centers regularly. Phlebotomy (bloodletting), when appropriate, should only be done by ACHD experts.

- All kinds of surgery can present risks for the CHD patient. This includes minor and routine procedures. Special attention is needed with anesthesia and to prevent heart infections (endocarditis). Patients on blood thinners also require special care. Ideally all surgery on these patients should be done at ACHD centers.

- Cyanotic patients must have their IV lines kept free of air bubbles.

Contraception and Pregnancy

- Women with CHD need to understand their risks from pregnancy. They should also understand any risks to their baby. All female ACHD patients should be educated about their birth control choices.

- Sometimes sterilization is considered due to high risk. Patients should be given all the information they need to make this decision. This includes the possible risks of the sterilization procedure. Information about future advances that might make pregnancy possible should also be included.

- Most female ACHD patients can have successful pregnancies. Those at higher risk should be seen at a center that specializes in high-risk pregnancy.

- All ACHD patients should be offered genetic counseling. The chance of having a baby with CHD varies widely based on defect. All women with CHD should be offered fetal echocardiography between their 16th and 18th week of pregnancy.

Exercise

- Studies on children suggest that exercise can be helpful for individuals with CHD. Exercise also benefits adults living with many other kinds of heart problems.

- Studies suggest that most ACHD patients have reduced ability to exercise. This includes patients with repaired simple defects.

- More studies are needed on the role of exercise in ACHD patients.

Psychosocial Issues

- Adults with complex CHD can face many challenges to their mental health. Some live with the knowledge that they face a significant risk of dying young. Their health can limit their education, social life, activity level, and/or work life. Some face challenges with their appearance, such as scarring, small body size, blue lips, and finger clubbing.

- CHD can affect lifestyle decisions, such as whether or not to leave home or marry. Some people may not have the information they need to make good choices in these areas. It is often difficult

for people to discuss their CHD with their friends and family. Counseling should be available to help people address these issues.

- More studies should be done on the emotional health of adults with CHD. Depression and anxiety have been shown to raise death rates in patients with other forms of heart disease.

- Individuals with CHD may have had bad experiences with medical care. As children some experienced fear and untreated pain. These experiences can affect their ability to cope in a medical setting. Resources should be offered to help address these issues. These should include education on pain management.

- Taking care of the emotional health of adults with CHD should be a priority.

Task Force 3: Workforce Description and Educational Requirements for the Care of Adults with Congenital Heart Disease

- Right now very few doctors have specific training in ACHD. Most ACHDers see adult cardiologists with little training in CHD. Others see pediatric cardiologists with little experience managing adult care.

- More programs should be started to train health care workers in all aspects of ACHD care. The use of physician assistants and nurse practitioners may help to increase ACHD services.

Task Force 4: Organization of Delivery Systems for Adults with Congenital Heart Disease

- Our health care system is not currently meeting the needs of adults living with heart defects.

- Regional ACHD centers should be established. There should be one regional ACHD center for every 5 to 10 million people. This means 30 to 50 ACHD centers should be developed throughout the US.

- Every adult with CHD should be evaluated at least once at an ACHD center. Ideally this includes those with “simple” heart defects (see chart).

- Individuals with more complex defects should be seen regularly at an ACHD center. Frequency of follow up depends on their specific condition.

- The role of the regional ACHD will vary based on the patient’s needs. ACHD patients may have both a local cardiologist and ACHD specialists. The patient should also have a primary care doctor. The ACHD center should provide expert advice and information to the other health care providers.

- A regional ACHD center should offer a team of specialized staff. This should include an ACHD cardiologist, other cardiologists, nurse practitioners and/or physician’s assistants, congenital heart surgeons and others. The team should work together to care for the patients.

- Regional ACHD Centers should provide:

    - Patient counseling
    - Operating rooms—Specially equipped with personnel familiar with CHD surgery, fully trained congenital cardiac perfusionists as well as a full array of diagnostic equipment.
    - Cardiac surgery—At least 2 congenital heart surgeons to provide 24 hour a day coverage.
    - Cardiac anesthesia

- Intensive Care
- In-patient service
- Transplantation
- Catheterization laboratory with staff specifically trained in the needs of ACHD patients.
- Noninvasive imaging service 24 hours a day, 7 days a week
- Electrophysiology service
- High-risk obstetrics
- Cardiac pathology

Task Force 5: Adults With Congenital Heart Disease: Access to Care

- Many ACHD patients have difficulty obtaining health insurance. Getting life insurance can also be a problem. Cardiologists should be willing to write letters describing conditions and long-term expectations.

- Insurance companies need to be made aware of the special needs of ACHD patients.

- Organized, effective and passionate advocacy for ACHD patients is needed. Patient groups and health care providers can work together for change.

The full text of the 32nd Bethesda Guidelines can be found online at http://www.acc.org/clinical/bethesda/beth32/dirindex.htm and in print: *Care of the Adult With Congenital Heart Disease. Presented at the 32nd Bethesda Conference, Bethesda, Maryland, October 2–3, 2000. J Am Coll Cardiol 2001; 37: 1161–98.* All references in this document can be found in the 32nd Bethesda Guidelines.

This document was prepared by the Medical Advisory Committee of the Adult Congenital Heart Association (ACHA), and reviewed by members of ACHA's Medical Advisory Board. All comments should be directed to info@achaheart.org.