Journal of the American College of Cardiology
Vol. 37, No. 5, 2001

ISSN 0735-1097/01/$20.00
Published by Elsevier Science Inc.
PII S0735-1097(01)01272-4

32nd Bethesda Conference:

"Care of the Adult With Congenital Heart Disease"
October 2–3, 2000

Journal of the American College of Cardiology
© 2001 by the American College of Cardiology
Published by Elsevier Science Inc.

Vol. 37, No. 5, 2001
ISSN 0735-1097/01/$20.00
PII S0735-1097(01)01272-4

BETHESDA CONFERENCE REPORT

32nd Bethesda Conference: "Care of the Adult With Congenital Heart Disease"*

Gary D. Webb, MD, FACC, *Conference Co-Chair*
Roberta G. Williams, MD, FACC, *Conference Co-Chair*

This Conference, sponsored by the American College of Cardiology, was held at Heart House, Bethesda, Maryland, October 2–3, 2000.

Participants

HUGH D. ALLEN, MD, FACC
Chief, Pediatric Cardiology
Physician-in-Chief
Columbus Children's Hospital
700 Children's Drive
Room ED622
Columbus, OH 43205-2696

JOSEPH S. ALPERT, MD, FACC
Professor of Medicine
Head of Department of Medicine
University of Arizona
Health Sciences Center
1501 N. Campbell Avenue
Tucson, AZ 85724-5035

JACQUES I. BENISTY, MD, FACC
Boston Adult Congenital Heart Service
Department of Cardiology
Children's Hospital
300 Longwood Avenue
Boston, MA 02115-5737

YVONNE BERNARD, RN
Case Manager
Division of Pediatric Cardiology
Vanderbilt University Medical Center
D2220 Medical Center North
Nashville, TN 37232-2572

ALAN C. BRAVERMAN, MD, FACC
Associate Professor of Medicine
One Barnes-Jewish Hospital Plaza
Suite 16419 East Pavilion
St. Louis, MO 63110
National Marfan Foundation

JOSEPH R. CARVER, MD, FACC
Senior Medical Director
Aetna U.S. Healthcare
980 Jolly Road
Mail Stop U21A
Blue Bell, PA 19422

MELVIN D. CHEITLIN, MD, MACC
Emeritus Professor of Medicine
University of California at San Francisco
San Francisco General Hospital, Room 5G1
1001 Potrero Avenue
San Francisco, CA 94110

JOHN S. CHILD, MD, FACC
Professor of Medicine
UCLA School of Medicine
Division of Cardiology
Department of Medicine
Los Angeles, CA 90024

RUTH L. COLLINS-NAKAI, MD, FACC
President, Canadian Cardiovascular Society
15239 43rd Avenue
Edmonton, Alberta T6G 5R3
CANADA

GORDON K. DANIELSON, MD, FACC
Roberts Professor of Surgery
Mayo Medical Center
200 First Street, SW
Rochester, MN 55905-0001

WILLIAM R. DAVIDSON, JR., MD, FACC
Director, Program for Adults with CHD
Director, Echocardiography
Cardiology H047
M.S. Hershey Medical Center
500 University Drive
Hershey, PA 17033

JOHN E. DEANFIELD, MD, FACC
Professor of Cardiology
Cardiothoracic Unit
Great Ormond Street Hospital
Institute of Child Health
Great Ormond Street
London, WC1N 3JH
UNITED KINGDOM

RICHARD M. DONNER, MD
Staff Pediatric Cardiologist
The Children's Hospital of Philadelphia
Division of Cardiology
34th and Civic Center Blvd
Philadelphia, PA 19104-4399

ANNIE DORE, MD, FRCP
Assistant Professor of Medicine
Montreal Heart Institute
5000 Belanger Est
Montreal, Quebec H1T 1C8
CANADA

DAVID J. DRISCOLL, MD, FACC
Professor of Pediatrics
Head, Section of Pediatric Cardiology
Mayo Clinic
200 First Street, S.W.
Rochester, MN 55905-0001

JOSEPH P. DROZDA, JR., MD, FACC
Principal, Carevergence
Associate Director, Unity Health Research
36 Picardy Hill Drive
Chesterfield, MO 63017-7127

SUSAN M. FERNANDES, MHP, PA-C
Boston Adult Congenital Heart Service
Department of Cardiology
Children's Hospital
300 Longwood Avenue
Boston, MA 02115-5737

*The recommendations set forth in this report are those of the Conference participants and do not necessarily reflect the official position of the American College of Cardiology.

When citing this document, the American College of Cardiology would appreciate the following citation format: 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.

This document is available on the American College of Cardiology Web site at www.acc.org. Reprints of this document are available for $5.00 each by calling 800-253-4636 (U.S. only) or by writing the Resource Center, American College of Cardiology, 9111 Old Georgetown Road, Bethesda, Maryland 20814.

ELYSE FOSTER, MD, FACC
Associate Professor of Clinical Medicine
University of California, San Francisco
Box 0214, Rm. 314A
Moffitt Hospital
505 Parnassus Avenue, M314A
San Francisco, CA 94143-0214

JAMES M. FOX, MS, MD
Cardiology Fellow
Division of Cardiology
University of Iowa Hospital
200 Hawkins Drive
Room E318-5
Iowa City, IA 52242

MICHAEL D. FREED, MD, FACC
Senior Associate
Department of Cardiology
Children's Hospital
300 Longwood Avenue
Boston, MA 02115-5737

VALENTIN FUSTER, MD, PHD, FACC
Director
Zena & Michael A. Wiener Cardiovascular Institute
Mount Sinai Medical Center
One Gustave Levy Place
Box 1030
New York, NY 10029-6500

ARTHUR GARSON, JR., MD, MPH, MACC
Senior Vice President
Dean for Academic Operations
Texas Children's Hospital
Baylor College of Medicine
6621 Fannin Street
MC1-4460
Houston, TX 77030

THOMAS P. GRAHAM, JR., MD, FACC
Director, Pediatric Cardiology
Vanderbilt Medical Center
1161 21st Avenue, South
(D2220 MCN)
Nashville, TN 37232-0001

LOUISE HARRIS, MD, CHB, FACC
Associate Professor of Medicine & Consultant Electrophysiologist
University of Toronto Congenital Cardiac Centre for Adults
Toronto General Hospital
150 Gerrard Street West
Gerrard Wing 3-562
Toronto, Ontario M5G 2C4
CANADA

JULIEN I. E. HOFFMAN, MD, FACC
Professor of Pediatrics (Emeritus)
Room M 1331, Box 0544
University of California
505 Parnassus Avenue
San Francisco, CA 94143-0001

CHARLES E. IRWIN, JR., MD
Professor of Pediatrics
Vice Chair, Department of Pediatrics
Director, Division of Adolescent Medicine
University of California, San Francisco
3333 California Street, Suite 245
Box 0503, LH 245
San Francisco, CA 94143-0503

JOHN A. JARCHO, MD, FACC
Medical Co-Director, Cardiac Transplant Program
Brigham & Women's Hospital
Cardiovascular Division
75 Francis Street
Boston, MA 02115

SAMUEL KAPLAN, MD, FACC
Professor of Pediatrics
University of California at Los Angeles
Heart Center, Suite 330
200 UCLA Medical Plaza
Los Angeles, CA 90095-6946

RAE-ELLEN W. KAVEY, MD, FACC
SUNY Health Science Center at Syracuse
Division of Pediatric Cardiology
725 Irving Avenue, Room 804
Syracuse, NY 13210
American Heart Association

HARLAN M. KRUMHOLZ, MD, FACC
Associate Professor of Medicine & Epidemiology and Public Health
Yale University School of Medicine
333 Cedar Street
Room IE-61 SHM
New Haven, CT 06520

MICHAEL J. LANDZBERG, MD, FACC
Director, Boston Adult Congenital Heart Group
Department of Cardiology
Children's Hospital & Brigham & Women's Hospital
300 Longwood Avenue
Boston, MA 02115-5737

JAMES LANGILL
66 Waverly Road
Woburn, MA 01801

RICHARD R. LIBERTHSON, MD
8 Hawthorne Place
Suite 110
Boston, MA 02114

ARIANE MARELLI, MD, FRCP(C), FACC
Assistant Professor of Medicine
McGill University
McGill University Health Care Center
1650 Cedar Avenue
Division of Cardiology E-500
Montreal, Quebec H3G 1A4
CANADA

JOHN E. MAYER, JR., MD, FACC
Professor of Surgery
Harvard Medical School
Sr. Associate in Cardiac Surgery
Children's Hospital
Cardiovascular Surgery
300 Longwood Avenue
Boston, MA 02115-5737

FRANK X. MCGOWAN, JR., MD
Associate Professor of Anesthesiology (Pediatrics)
Co-Director, Cardiac Anesthesia Services
Cardiac Anesthesia Service
Children's Hospital
300 Longwood Avenue
Boston, MA 02115-5737

PETER MCLAUGHLIN, MD, FACC
Professor of Medicine
University of Toronto Congenital Cardiac Centre for Adults
University Health Network
Toronto General Hospital
200 Elizabeth Street
Eaton North, 12-224
Toronto, Ontario M5G 2C4
CANADA

ROGER B. B. MEE, MD, CHB
Chairman, Pediatric and Congenital Heart Surgery
The Cleveland Clinic Foundation
9500 Euclid Avenue, M-41
Cleveland, OH 44195-0001

PAMELA D. MINER, RN, MN, NP
Nurse Practitioner
Ahmanson/UCLA Adult Congenital Heart Disease Center
10833 LeConte Avenue
Room 47-123 CHS
Los Angeles, CA 90095

DOUGLAS S. MOODIE, MD, FACC
Chairman, Division of Pediatrics
The Cleveland Clinic Foundation
9500 Euclid Ave., A120
Cleveland, OH 44195-0001

JOHN W. MOORE, MD, MPH, FACC
The Heart Center for Children
St. Christopher's Hospital for Children
Erie Avenue at Front Street
Philadelphia, PA 19134
American Academy of Pediatrics

DANIEL J. MURPHY, JR., MD, FACC
The Cleveland Clinic Foundation
9500 Euclid Avenue, M-41
Cleveland, OH 44195-0001
International Society for Adult Congenital Cardiac Disease

SUZANNE R. PATTEE, JD
Vice President of Public Policy & Patient Affairs
Cystic Fibrosis Foundation
6931 Arlington Road, #200
Bethesda, MD 20814
Cystic Fibrosis Foundation

GAIL D. PEARSON, MD, ScD, FACC
Leader, Heart Development, Function & Failure
Scientific Research Group
Heart Research Program
National Heart, Lung & Blood Institute
National Institutes of Health
6701 Rockledge Drive
Room 9200, MSC 7940
Bethesda, MD 20892-7940
National Heart, Lung & Blood Institute

JOSEPH K. PERLOFF, MD, FACC
Director, Ahmanson/UCLA Adult Congenital Heart Disease Center
Professor of Medicine and Pediatrics
University of California at Los Angeles
Division of Cardiology
Room 47-123 CHS
10833 LeConte Avenue
Los Angeles, CA 90095-1679

REED E. PYERITZ, MD, PhD
Chair, Department of Human Genetics
Professor of Human Genetics, Medicine & Pediatrics
MCP Hahnemann School of Medicine
320 E North Avenue
Pittsburgh, PA 15212-4772

GRAHAM J. REID, PhD, C Psych
Assistant Professor, Psychology and Family Medicine
University of Western Ontario
Asst. Professor, Public Health Sciences & Medicine
University of Toronto
SSC 7310
London, Ontario N6A 5C2
CANADA

JOHN G. REISS, PhD
Director, Policy and Program Affairs
Associate Professor of Pediatrics
University of Florida
5700 SW 34th Street, Suite 323
Gainesville, FL 32608
Institute for Child Health Policy

SARA ROSENBAUM, JD
Hirsh Professor, Health Law & Policy
George Washington University Medical Center
School of Public Health and Health Services
2021 K Street, NW
Suite 800
Washington, DC 20006

ISOBEL A. RUSSELL, MD, PhD
Associate Professor of Anesthesia
Dept. of Anesthesia & Perioperative Care
University of California at San Francisco
521 Parnassus Avenue
San Francisco, CA 94143-0648

DAVID J. SAHN, MD, FACC
Professor and Director, Clinical Care Center for Congenital Heart Disease
Pediatric Cardiology
Oregon Health Sciences University
3181 SW Sam Jackson Park Road
Portland, OR 97201-3098

MATHEW SERMER, MD
Chief of Obstetrics
Toronto General Hospital
200 Elizabeth Street
Toronto, Ontario M5G 2C4
CANADA

SAMUEL C. SIU, MD, SM, FACC
Director of Research
University of Toronto Congenital Cardiac Centre for Adults
Division of Cardiology, PMCC 3-526
200 Elizabeth Street
Toronto, Ontario M5G 2C4
CANADA

DAVID J. SKORTON, MD, FACC
Vice President for Research
The University of Iowa
201 Gilmore Hall
Iowa City, IA 52242-1320

LAURENCE J. SLOSS, MD
Co-Director
Boston Adult Congenital Heart Service
Department of Cardiology
Children's Hospital
Department of Cardiology
300 Longwood Avenue
Boston, MA 02115-5737

JANE SOMERVILLE, MD, FACC
Professor
Royal Brompton Hospital
81/83 Harley Street
London, W1N 1DE
UNITED KINGDOM

DAVID M. STRASSER, MD, FACC
Medicor Associates, Inc.
104 E. 2nd Street
Erie, PA 16507

JON B. TINGELSTAD, MD, FACC
American Board of Pediatrics
111 Silver Cedar Court
Chapel Hill, NC 27514
American Board of Pediatrics

SUSIE C. TRUESDELL, PA, MBA
Xerox Corporation
80 Linden Oaks Parkway
Mail Stop 0804-02A
Rochester, NY 14625

KAREN UZARK, PhD, PNP
Director of Cardiology Process and Improvement and Clinical Outcomes
Children's Hospital Medical Center
Cardiology Department
3333 Burnet Avenue, OSB-4
Cincinnati, OH 45229-3039

GEORGE F. VAN HARE, MD, FACC
Associate Professor of Pediatrics
Stanford University
750 Welch Road, Suite 305
Palo Alto, CA 94304

ROBERT A. VOGEL, MD, FACC
Herbert Berger Professor of Medicine
Head, Division of Cardiology
University of Maryland Hospital
22 S. Greene Street, Rm. S3B06
Baltimore, MD 21201-1544

CAROLE A. WARNES, MD, FACC
Professor of Medicine
Mayo Clinic
Cardiovascular Diseases
Rochester, MN 55905

GARY D. WEBB, MD, FACC
Director
University of Toronto Congenital Cardiac Centre for Adults
Toronto General Hospital
200 Elizabeth Street
12EN 215
Toronto, Ontario M5G 2C4
CANADA
President—Canadian Adult Congenital Heart Network
Conference Co-Chair

ROBERTA G. WILLIAMS, MD, FACC
Chairman, Department of Pediatrics
KECK School of Medicine
University of Southern California
Vice President for Pediatrics and Academic Affairs
Childrens Hospital Los Angeles
4650 Sunset Blvd., MS 71
Los Angeles, CA 90027
Conference Co-Chair

Conference Steering Committee

GARY D. WEBB, MD, FACC, Conference Co-Chair
ROBERTA G. WILLIAMS, MD, FACC, Conference Co-Chair
JOSEPH S. ALPERT, MD, FACC
JOHN S. CHILD, MD, FACC
RUTH L. COLLINS-NAKAI, MD, FACC
ELYSE FOSTER, MD, FACC
ARTHUR GARSON, Jr., MD, MPH, MACC
THOMAS P. GRAHAM, Jr., MD, FACC
MICHAEL J. LANDZBERG, MD, FACC
RICHARD LIBERTHSON, MD
DANIEL J. MURPHY, Jr., MD, FACC
DAVID J. SKORTON, MD, FACC
CAROLE A. WARNES, MD, FACC

Staff

American College of Cardiology

CHRISTINE W. McENTEE, Executive Vice President
MARIE E. MICHNICH, DrPH., Senior Associate Executive Vice President
CHARLENE L. MAY, Director, Document Development and Practice Guidelines
LISA B. BRADFIELD, Manager, Document Development

Journal of the American College of Cardiology
© 2001 by the American College of Cardiology
Published by Elsevier Science Inc.

Vol. 37, No. 5, 2001
ISSN 0735-1097/01/$20.00
PII S0735-1097(01)01272-4

32ND BETHESDA CONFERENCE

Care of the Adult With Congenital Heart Disease: Introduction

Gary D. Webb, MD, FACC, *Conference Co-Chair,*
Roberta G. Williams, MD, FACC, *Conference Co-Chair*

Consider the changes in the practice of cardiology in the lifetime of a 40-year-old patient with tetralogy of Fallot. In the 1960s it was common for adult and pediatric cardiologists to practice together in a medical school setting, with the exception of a few freestanding children's hospitals. Patients were discussed at a joint conference. Without the demands of highly technical procedures, there was a greater commonality of knowledge and skills than we find today. Infant surgery was practically non-existent, and surgery on young children was usually confined to palliative procedures. Repair of lesions such as tetralogy of Fallot or atrial septal defect was generally delayed until early adolescence. Patients who survived until adult life were few.

The rapid technological developments that have occurred in the three past decades have enabled patients with more complex congenital cardiac defects to survive into adult life. Now, for the first time, the number of adults with congenital heart disease (CHD) equals the number of children with the disorder. The range of abnormalities and the complexities of postoperative anatomy are now well beyond the educational and experiential background of the adult cardiologist. Pediatric cardiologists, who often follow their patients into adult life, are at a disadvantage when confronted by superimposed acquired adult diseases. Both groups of subspecialists are drawn deeper into the complexities of their own fields and have vanishingly few opportunities to interact in an ongoing professional forum. A few adult congenital heart disease (ACHD) programs have served as referral centers for adult patients who require surgical or catheter intervention, but most patients do not have their cases reviewed in these centers. There is no organized effort at monitoring clinical outcomes for these patients, and the frequency with which these patients appear late at tertiary centers with avoidable complications indicates that health care delivery to this population falls far short of that for adults with acquired heart disease or children with CHD.

The transition from childhood to adulthood is particularly difficult for patients with chronic disease. Some are multiply-handicapped with cognitive and physical disabilities. The less disabled do not qualify for public insurance and are at risk for the development of secondary disability as a result of inadequate continuity of care. Adolescence is a time of particular anxiety about conformity, social success, and uncertainty about the future. Denial is a frequently used defense, coupled with a sense of immortality and a desire for risk taking. Relocation for educational or occupational reasons is common and often results in the transfer of inadequate information to the new provider. When one is distracted by the demands of job and family, it is easier to put aside routine health care, particularly when one is asymptomatic. Considering the human and financial resources that have been expended in nurturing these individuals to adult age, it is important to improve the system of care for adults with CHD in order for them to maintain the functional status that has been so hard won.

The American College of Cardiology is the professional organization where adult cardiovascular specialists, pediatric cardiologists, and cardiothoracic surgeons join together in the interests of care delivery, professional education, and advocacy. The authors are grateful for the opportunity to bring together these formidable resources in the format of the Bethesda Conference. The conference was organized into five sections that represent: 1) the present number of adults with CHD, 2) the special needs of these patients, 3) the workforce needed to deliver ACHD health care, 4) recommendations for a system of health care delivery that would produce the best clinical outcomes with the most efficient use of resources, and 5) the steps needed to ensure access to adequate health care for these patients. The answers reduce to three requirements: 1) education of a modest number of ACHD specialists who can lead highly specialized teams and serve as a referral and continuing educational resource for the greater number of providers with training in either adult or pediatric cardiology; 2) integration of highly specialized centers with community-based providers and with each other in order to provide a high level of care for all patients and to provide an informational base for continual improvement in care; and 3) unfettered access to an appropriately trained provider for all adults with CHD. These actions would require a modest outlay of resources, but the status quo will result in greater long-term expenditures for terminal care and loss of productivity for this ever-growing population. The product of this conference is a comprehensive analysis of the problem and a recommendation for corrective steps. The realization of the vision articulated in the following sections will require the passion and constancy of focus that have been exhibited by so many of the participants in this conference.

Journal of the American College of Cardiology
© 2001 by the American College of Cardiology
Published by Elsevier Science Inc.

Vol. 37, No. 5, 2001
ISSN 0735-1097/01/$20.00
PII S0735-1097(01)01272-4

Summary of Recommendations—Care of the Adult With Congenital Heart Disease

As a result of extremely successful diagnostic and treatment strategies developed and employed over the past 40 years, the number of adults with congenital heart defects in the U.S. has reached ~800,000. Of these, half are complex enough to require ongoing follow-up and treatment by health care professionals with expertise in the care of these patients. The health care system in the U.S. has developed neither a plan nor the required systems and facilities to care for these patients. Consequently, for the present and foreseeable future, most cardiologists who treat adults with congenital heart disease (CHD) have had informal training and considerable experience in the care of these patients. Over the next 10 years, more specifically trained adult congenital heart disease (ACHD) cardiologists should be trained in this subspecialty.

The American College of Cardiology (ACC) convened this 32nd Bethesda Conference to study the needs of these patients and to invite expert participants to recommend changes that will improve these patients' care and access to the health care system.

The Conference report consists of an introduction and five Task Force reports. These documents largely focus on the coming decade. A series of recommendations are made, as summarized here.

ORGANIZATION OF CARE

- The care of adults with CHD should be coordinated by regional ACHD centers.
- One regional ACHD center should be created to serve a population of 5 million to 10 million people. Approximately 30 to 50 such centers should be developed or strengthened across the entire U.S.
- Adults with moderate and complex CHD (defined in the Task Force 1 report) will require regular evaluations at a regional ACHD center and will benefit from maintaining contact with a primary care physician.
- The ACC should recommend to the National Heart, Lung and Blood Institute (NHLBI) and/or the Agency for Health Care Research and Quality (AHRQ), the formation of adult congenital cardiac centers for documenting and improving outcomes, education, and research.
- Each pediatric cardiology program should identify the ACHD center where their patients will be transferred.
- Every adult cardiology and adult cardiac surgical center, as well as every cardiologist, should have a referral relationship with a regional ACHD center.
- All emergency care facilities should have an affiliation with a regional ACHD center.
- Physicians without specific training and expertise in ACHD should manage adults with moderate and complex CHD only in collaboration with physicians with advanced training and experience in the care of adults with CHD.
- An ACHD cardiologist should evaluate all adults with moderate and complex CHD at least once. The initial ACHD evaluation allows stratification of these patients according to risk and anticipated management difficulty.
- Patients with moderate or complex CHD usually require hospital admission or transfer to a regional ACHD center for urgent or acute care.
- Most cardiac catheterization and electrophysiology procedures for adults with moderate and complex CHD should be performed in regional ACHD centers with appropriate experience in CHD and in laboratories with appropriate personnel and equipment. After consultation with staff in regional ACHD centers, it may be appropriate for local centers to perform these procedures.
- Cardiovascular surgical procedures in adults with moderate and complex CHD should generally be performed in regional ACHD centers with specific experience in the surgical care of these patients.
- Each regional ACHD center should participate in a medical and surgical database aimed at defining and improving outcomes in adults with CHD.
- Appropriate clinical records for each patient should be kept in the regional ACHD center; the primary care provider and patient should also keep such records.

WORKFORCE DESCRIPTION AND EDUCATIONAL REQUIREMENTS

- A joint task force of the American Board of Internal Medicine and the American Board of Pediatrics, facilitated by the ACC, should be formed to determine the specific pathways and years of training required for Level 3 ACHD subspecialist cardiologists.
- Level 3 training programs should collaborate to maximize learning opportunities for the ACHD cardiologists-in-training and provide continuing education for trainees, graduates, and ACHD practitioners.
- A network of centers of research and education in ACHD should be created and funded through the NHLBI.
- Research fellowships in ACHD should be created so that these fellows can dedicate 75% to 100% of their time in protected research over a two- to three-year period.
- Training programs for other key staff (e.g., nurses, physician assistants, psychologists, social workers, other nonphysician personnel) on ACHD teams should be established.

- The ACC should lobby Congress for an educational loan repayment program for ACHD specialists to lessen the financial constraint of the prolonged educational process leading to an academic career.

ACCESS TO CARE

The ACC should:

- Work with the American Heart Association (AHA) to develop a strategic plan for an organized advocacy group, which includes health care professionals and patients and their families in the context of a public relations campaign.
- Collaborate with the AHA to develop educational materials to guide adolescent and adult patients in the transition to independence, including the need for health (and perhaps life) insurance, barriers that may exist in obtaining coverage, and strategies to obtain optimal coverage.
- Sponsor a multicenter study with economic forecasting to develop a better understanding of the true economic impact (e.g., payments, future income potential) of CHD in the adult.
- Include, in formal and regular discussions with insurance companies and other public and private payers and purchasers, information on the special problems encountered and expertise necessary in the care of adolescents and adults with CHD.
- Reduce barriers to multidisciplinary services by developing innovative reimbursement methods. Pilot programs established between one or more ACHD centers and major payers (public and private) should be encouraged.
- Work at the chapter level with state legislators to specify CHD in a demonstration project of the Work Incentives Improvement Act.
- Recommend that physicians discuss individual patient coverage concerns with insurance company medical directors.
- Advocate health care coverage for all. As an incremental step, all adults with CHD should be covered, thus removing a significant barrier to access.
- Develop additional educational materials to help adolescent and adult patients as they approach the job market, focusing on their legal rights (e.g., health should not be discussed during an interview), tips for success, and where to go for job training and vocational counseling.
- Recommend, at the patient's request, that individual physicians work directly with patients, their schools, and their employers or potential employers to optimize opportunities.

SPECIAL NEEDS OF ADULT PATIENTS WITH CHD

Each patient's transition to adult life should include:

- A structured plan to help patients transition from pediatric to adult CHD care.
- Individual patient education regarding their diagnosis and specific health behaviors
- A "health care passport." Important historic information, including comprehensive diagnostic data, procedures, operations, and medications, should be kept by patients as a summary of past and present important health issues. The ACC should support the development of a health care passport, which would be useful for all patients with CHD and their health care providers.
- A continuum of vocational assessment, beginning in late childhood and proceeding through the adolescent and young adult years.
- Transfer of information to the patient and family, within a transition program, at a rate commensurate with the prevailing psychosocial development and circumstances of the patient.

Recommendations Regarding Noncardiac Surgery

- Noncardiac operations on patients with moderate and complex CHD should be performed at a regional ACHD center, with the consultation of an anesthesiologist with experience in CHD. This applies particularly when more complex surgery is indicated, or when patients have adverse risk factors, including poor functional class, pulmonary hypertension, CHF, and cyanosis.

Recommendations Regarding Reproductive Issues

- Contraceptive counseling must be available, when appropriate, to all patients with CHD.
- A multidisciplinary team at a regional ACHD center is needed for pregnancy care and delivery, as well as the management of indeterminate, intermediate-, or high-risk patients.
- The recurrence risk of CHD is highly variable and should be discussed prospectively with all patients. Genetic counseling should be made available through regional ACHD centers.

Recommendations Regarding Exercise and Rehabilitation

- Guidance for athletic participation for patients with CHD can be found in the published recommendations of the 26th Bethesda Conference report, which represents the best consensus data available.
- The efficacy and safety of exercise rehabilitation programs for adults with CHD have not been studied, and research in this area should be supported.

Recommendations Regarding Psychosocial Issues

- The emotional health of adults with CHD should be a priority in the overall care of this patient population. Appropriate screening and referral sources for treatment should be available at all regional ACHD centers.
- Tools for screening for psychosocial problems in this population should be developed and tested.

- Data should be developed to assess the effectiveness of regular follow-up care on the long-term physical and psychosocial health of adults with CHD.
- Professionals and facilities for the treatment of psychological disorders are scarce, and creative solutions for counseling patients in groups or those who live a far distance away should be developed.

CONCLUSIONS

The participants in this Bethesda Conference on adults with CHD have compiled these recommendations, along with supporting information, to construct a road map for future actions. Action will be needed from governments, health insurance organizations, health care institutions, clinical and academic units, and health care providers.

Ongoing efforts made by the ACC, as well as the present and future leaders, in the care of these patients will be required to help realize this vision over the next decade. We urge readers to acknowledge the serious problems in current health care delivery to these patients in the U.S. and commit themselves to doing what they can to become a part of the solution, which requires the collaboration of many individuals and organizations across this country.

Journal of the American College of Cardiology
© 2001 by the American College of Cardiology
Published by Elsevier Science Inc.

Vol. 37, No. 5, 2001
ISSN 0735-1097/01/$20.00
PII S0735-1097(01)01272-4

TASK FORCES

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

Carole A. Warnes, MD, FACC, *Co-Chair,* Richard Liberthson, MD, *Co-Chair,*
Gordon K. Danielson, Jr, MD, FACC, Annie Dore, MD, FRCP, Louise Harris, MD, ChB, FACC,
Julien I. E. Hoffman, MD, FACC, Jane Somerville, MD, FACC, Roberta G. Williams, MD, FACC,
Gary D. Webb, MD, FACC

The extraordinary advances in cardiac surgery, intensive care, and noninvasive diagnosis over the last 50 years have led to an enormous growth in the U.S. and throughout the world in the number of adults with congenital heart disease (CHD). Approximately 85% of babies born with cardiovascular anomalies can expect to reach adulthood, and with continued improvement in surgical technique, this could increase further in the next two decades (1). In Canada, it is estimated that the number of survivors with adult congenital heart disease (ACHD) will increase from 94,000 in 1996 to 124,000 by the end of 2006. Although there is a general recognition that there are large numbers of adults with CHD in the U.S., accurate statistics are lacking. Reported prevalence rates of CHD in newborns vary widely and depend, to some extent, on lesion inclusion and exclusion criteria. For example, some studies include ventricular septal defects (VSDs); however, about two thirds of these individuals no longer have a VSD by adult age. Many studies exclude bicuspid aortic valves, which are present in 1% of live births. In addition, different methods of ascertainment (e.g., physical examination, echocardiography, registry data) yield varying prevalence rates of CHD in infancy.

A recent English study (2) reviewed all births in one health region (Newcastle) between 1985 and 1994, and noted 1,942 cases of CHD in a population of 377,310 live births (incidence of 5.2 per 1,000). Of these newborns, 1,514 were predicted to survive ≥16 years. Because additional diagnoses are sometimes made later in childhood, at least 2,192 children were expected to survive ≥16 years. Also, an estimated 784 would require follow-up in adult life. These figures predict the need for follow-up of adults with CHD, for >200 cases per 100,000 live births, or >1,600 cases every year in the U.K. (assuming a population of 50 million). Assuming a population of 280 million in the U.S., that would mean an increase of 8,960 adult cases annually, or 89,600 cases in the current decade.

Most studies from the mid 1980s onward, however, as well as more recent Canadian studies, report the number of CHD births to be close to 10 in 1,000 live births (3). Defining the exact size and composition of this population in adulthood is challenging, because data are lacking. An important mandate of this Bethesda Conference is to estimate patient numbers, which are essential for program planning and resource allocation. On the basis of the U.S. census data, the documented birth rates from 1940 to 1989 were averaged (Tables 1–3). The diagnoses corresponding to complex, moderate, and mild lesions are shown in Tables 4 through 6, and are those used by Task Force 4. Based on a documented incidence of 1.5 in 1,000 live births for complex CHD (Table 1) and by extrapolating likely survival rates for the early through more recent years, the approximate numbers of survivors in this group were derived. The incidence of 1.5 in 1,000 live births was based on the large New England Regional Infant Cardiac Program (NERICP) review of catheterization data, surgical findings, and postmortem diagnoses (4). Using this approach, ~117,000 adults with truly complex CHD are estimated to live in the U.S. in the year 2000. With improved surgical techniques, this number can be anticipated to increase over the next decade.

Using a similar model, Table 2 demonstrates the anticipated survival, to the year 2000, of patients with moderate CHD, as defined in Table 5. A prevalence of 2.5 in 1,000 is derived from published data on children, as well as some patients who began with more simple lesions but acquired complications (e.g., VSD with valve lesions, patent ductus arteriosus causing left heart dilation) (Table 7). These estimates predict an adult population of 302,000 with moderate CHD by the year 2000 in the U.S.

Estimating the number of adult patients with simple CHD (Tables 3 and 6) is more difficult. To utilize the absolute prevalence of simple lesions detected in infancy would grossly overestimate the number of adult survivors, because most VSDs will have closed by adulthood, and these patients will no longer be considered to have CHD. Thus, there will be considerable "attrition" of the numbers of patients with VSDs between the incidence at birth and the prevalence in adulthood. Most patients with a patent ductus arteriosus will undergo surgical or spontaneous closure in childhood (by definition, therefore, remaining "simple"), but a small proportion will remain patent, many needing closure, and are therefore defined as "moderate" cases. By utilizing these assumptions (Table 7), the prevalence of these lesions is derived: ~2.2 in 1,000. Thus, the estimated survival of patients with simple CHD in the U.S. to the year 2000 is 368,800. A conservative estimate of the total number of survivors—combining the mild, moderate, and complex subgroups—is 787,800. The addition of those with isolated bicuspid aortic valves would dramatically

Table 1. Estimated Prevalence of Complex Congenital Heart Disease in the U.S.

Year of Birth	Birth Rate/ Years	Prevalence (1.5/1,000)	Survival Rate (First Year)	Survival Rate (to Year 2000)
1940–1959	3 million	4,500	20%	10%
	× 20 years	90,000	18,000	9,000
1960–1979	4 million	6,000	65%	50%
	× 20 years	120,000	78,000	60,000
1980–1989	4 million	6,000	85%	80%
	× 10 years	60,000	51,000	48,000
Totals			147,000	117,000

increase this number. The moderate and complex subgroups—totaling 419,000 patients—need periodic (e.g., every 6–24 months) follow-up in a regional ACHD center.

These figures may well be underestimates for two important reasons. First, they are based on the incidence of CHD presenting in infancy and childhood, but *at least* 10% of cases seen in an ACHD clinic (in particular, secundum atrial septal defect, Ebstein's anomaly, and congenitally corrected transposition) are not diagnosed until adulthood. In addition, increasing numbers of immigrants to the U.S. add to the patient population. Therefore, a conservative estimate of patients needing periodic follow-up at a regional ACHD center is ~400,000. Although these predictions, again, are based on several assumptions, they provide a framework to estimate current and future resource requirements necessary to provide optimal care.

This population growth is also reflected in the growth of individual regional ACHD centers. In Toronto, a 269% expansion in the outpatient work load was noted over a 10-year period between 1987 and 1997. Similarly, an increase in the number of admissions to a large ACHD unit in the U.K. is shown in Figure 1. Notably, these admissions continue to increase, particularly for patients >30 years of age; by 1996, 30% of patients admitted were >40 years of age.

DISEASE PATTERNS

Data on the basic diagnosis and age of outpatients in a large unit in the U.K. in 1997 are also presented (Figs. 2 and 3). Complex lesions, such as tricuspid atresia and single-ventricle physiology, are well represented in patients >20 years of age, particularly in those >30 years of age, in current ACHD centers. The age range of patients seen in two large clinics is shown in Figure 4; they were older in the Mayo Clinic than in the Toronto series, where 50% versus 30% of patients were ≥40 years of age. These more complex patients are obviously vulnerable to additional acquired co-morbidities that impact both their cardiac and medical care, including hypertension, pulmonary, renal, and myocardial disease, and coronary artery disease. It is estimated that ~55% of the adult patient population is at medium to high-risk (defined as those at significant risk for premature death, re-operation, and complications) and thus need to be seen regularly in ACHD regional centers and followed for life. These patients include those with atresia, single-ventricle physiology, transposition variants, Ebstein's anomaly, tetralogy of Fallot, pulmonary vascular disease, and complex septal defects. Periodic review at a regional ACHD center continues to offer advantages over a general cardiac evaluation, particularly regarding the timing and type of intervention, follow-up strategy, and general recommendations (5). Approximately 45% of patients with mild defects, such as a small VSD or mild pulmonary valve stenosis, will not require regular follow-up in a regional ACHD center, but might benefit from at least one review at such a center at the discretion of the patient's physician.

The profile of this patient population will change over the next few decades, not only because of advancing age, but also with improved survival of patients with complex anomalies. In addition, with the impetus to perform definitive repair at an earlier age and with changing operative procedures, there will be changes in the anticipated disease patterns. Many adult survivors will have different hemodynamic and cardiac problems from those currently seen. For example, an infant with transposition of the great arteries

Table 2. Estimated Prevalence of Moderate Congenital Heart Disease in the U.S.

Year of Birth	Birth Rate/ Years	Prevalence (2.5/1,000)	Survival Rate (First Year)	Survival Rate (to Year 2000)
1940–1959	3 million	7,500	60%	55%
	× 20 years	150,000	90,000	82,500
1960–1979	4 million	10,000	70%	65%
	× 20 years	200,000	140,000	130,000
1980–1989	4 million	10,000	90%	90%
	× 10 years	100,000	90,000	90,000
Totals			320,000	302,500

Table 3. Estimated Prevalence of Simple Congenital Heart Disease in the U.S.

Year of Birth	Birth Rate/ Years	Prevalence (2.2/1,000)	Survival Rate (First Year)	Survival Rate (to Year 2000)
1940–1959	3 million	6,600	95%	90%
	× 20 years	132,000	125,400	118,000
1960–1979	4 million	8,800	95%	95%
	× 20 years	176,000	167,200	167,200
1980–1989	4 million	8,800	95%	95%
	× 10 years	88,000	83,600	83,600
Totals			376,200	368,800

will no longer have a Mustard or Senning procedure (with its late problems of systemic ventricular dysfunction and arrhythmias), but might be anticipated to have an arterial switch procedure and encounter quite different cardiac sequelae in adult life. Patients with complex single-ventricle physiology and various modifications of the Fontan procedure will increase in number. Perhaps with refinements in noninvasive diagnosis and earlier definitive repair of shunt lesions, the prevalence of pulmonary vascular disease and Eisenmenger syndrome in the adult population could be expected to diminish. These patients with complex malformations are subject to more diverse and numerous late complications and must be seen regularly at a regional ACHD center, to which they should have direct access. They need more intensive follow-up and probably more frequent re-evaluations and interventions.

SPECIAL RESOURCES

Impact of cardiac surgery. In the largest congenital cardiac center in the U.K., one in five admissions was for cardiac surgery. The Society of Cardiothoracic Surgeons of the U.K. Registry for 1998/1999 reports that in the U.K., 3,836 congenital heart operations were performed, with a mortality rate of 4.7%. There were 339 patients ≥16 years of age, with a mortality rate of 2.1%, but the data were not stratified according to low- and high-volume units, nor were they audited.

Some centers reported a surprisingly low number of ACHD operations per year, although expertise is often focused in centers where the same surgeons operate on both pediatric and adult patients, so the numbers can be combined. Previously published data from Stark et al. (6) have shown that mortality is higher in centers with lower operative volume, highlighting the risk of performing the "occasional" operation on adult patients with CHD.

It is estimated in the U.S. that 20,000 operations for CHD are performed every year. Based on pediatric data, low-volume centers have a higher mortality. The outcome is likely to be worse for adult patients who do not always have the benefit of a surgeon with special expertise and training in CHD. It is important, both medically and financially, to concentrate resources and funding and place patients in specialized centers. A close collaboration is necessary between experienced and trained cardiologists, echocardio-

Table 4. Types of Adult Patients With Congenital Heart Disease of Great Complexity*

- Conduits, valved or nonvalved
- Cyanotic congenital heart (all forms)
- Double-outlet ventricle
- Eisenmenger syndrome
- Fontan procedure
- Mitral atresia
- Single ventricle (also called double inlet or outlet, common or primitive)
- Pulmonary atresia (all forms)
- Pulmonary vascular obstructive diseases
- Transposition of the great arteries
- Tricuspid atresia
- Truncus arteriosus/hemitruncus
- Other abnormalities of atrioventricular or ventriculoarterial connection not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

*These patients should be seen regularly at adult congenital heart disease centers. Modified from Connelly MS, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.

Table 5. Types of Adult Patients With Congenital Heart Disease of Moderate Severity*

- Aorto-left ventricular fistulae
- Anomalous pulmonary venous drainage, partial or total
- Atrioventricular canal defects (partial or complete)
- Coarctation of the aorta
- Ebstein's anomaly
- Infundibular right ventricular outflow obstruction of significance
- Ostium primum atrial septal defect
- Patent ductus arteriosus (not closed)
- Pulmonary valve regurgitation (moderate to severe)
- Pulmonic valve stenosis (moderate to severe)
- Sinus of Valsalva fistula/aneurysm
- Sinus venosus atrial septal defect
- Subvalvar or supravalvar aortic stenosis (except HOCM)
- Tetralogy of Fallot
- Ventricular septal defect with
  - Absent valve or valves
  - Aortic regurgitation
  - Coarctation of the aorta
  - Mitral disease
  - Right ventricular outflow tract obstruction
  - Straddling tricuspid/mitral valve
  - Subaortic stenosis

*These patients should be seen periodically at regional adult congenital heart disease centers. Modified from Connelly MS, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.
HOCM = hypertrophic obstructive cardiomyopathy.

Table 6. Types of Adult Patients With Simple Congenital Heart Disease*

Native disease
Isolated congenital aortic valve disease
Isolated congenital mitral valve disease (e.g., except parachute valve, cleft leaflet)
Isolated patent foramen ovale or small atrial septal defect
Isolated small ventricular septal defect (no associated lesions)
Mild pulmonic stenosis
Repaired conditions
Previously ligated or occluded ductus arteriosus
Repaired secundum or sinus venosus atrial septal defect without residua
Repaired ventricular septal defect without residua

*Those patients can usually be cared for in the general medical community.
Modified from Connelly MS, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.

graphers, interventional cardiologists, surgeons, and anesthesiologists, with well-trained nurses on every team. The expert surgical care provided to children with cardiac anomalies must also be provided to adults. Re-operations are frequent, and the overall mortality is higher in patients having a re-operation versus a first operation (7). In one U.S. center (Mayo Clinic) following >1,800 patients, 1,243 of whom had cardiac surgery, almost 50% had two or more operations and 290 (23%) had three or more operations. This necessity for re-operation (particularly in patients with bioprosthetic valves and extracardiac conduits), again emphasizes the need for special surgical expertise in CHD. The types of operations in adult patients in a single center (Mayo Clinic) by diagnosis and age are shown in Table 8.

Operative mortality varies according to the basic diagnosis, the type of surgical repair, and the complexity of the anatomy. Re-operation poses technical difficulties for the surgeon because of adhesions (especially between the heart, aorta or conduit, and sternum), lack of retrosternal space, loss of anatomic landmarks (especially the coronary arteries) or the development of collateral vessels. In addition, there may be deleterious effects of all previous bypass operations on long-term myocardial function. Cyanotic patients face a higher mortality and more postoperative complications.

Table 7. Estimated Prevalence of Simple Congenital Heart Lesions in Infancy, as Compared With Prevalence in Adulthood

	Prevalence in Infancy (per 1,000)	Estimated Prevalence of Simple Lesions in Adulthood (per 1,000)
VSD*	3	0.3
PDA†	0.6	0.5
ASD‡	0.9	0.6
PS§	0.6	0.5
AS	0.3	0.3
Totals	5.4	2.2

*Most ventricular septal defects (VSDs) are closed by adulthood and are no longer a problem (i.e., the patients no longer have a "lesion"). †Most patent ductus arteriosi (PDAs) close spontaneously or are closed in childhood, and therefore remain in the "simple" category. ‡Most atrial septal defects (ASDs) are closed in childhood, and therefore remain in the "simple" category. §Most pulmonary stenoses (PS) remain in the "simple" category; some will become moderate or severe; and some will develop pulmonary regurgitation, and therefore be defined as "moderately complex."
AS = aortic stenosis.

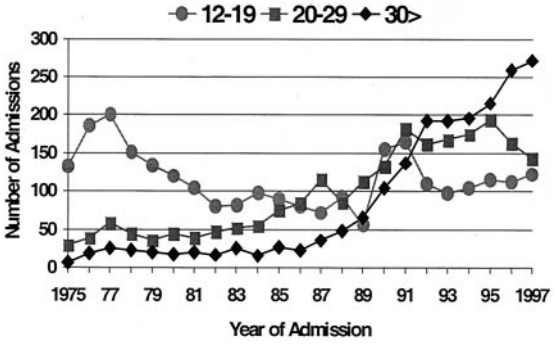

Figure 1. Age range of patients with CHD at hospital admission in a single center from 1975 onwards. The unit was opened as an adolescent unit in 1975 at the National Heart Hospital, joined by the Royal Brompton Hospital in 1990. Statistics from Jane Somerville, London, U.K.

Increasing age is associated with a higher mortality because additional co-morbid factors (as outlined previously) increase the operative risk. A detailed preoperative evaluation performed by an experienced medical and surgical team is essential. Transthoracic and transesophageal echocardiography, cardiac catheterization, and magnetic resonance imaging are necessary complementary tools to help the physicians make appropriate decisions. Holter monitoring and electrophysiologic study may determine if significant arrhythmias are present. Adults often report that they are asymptomatic as they adapt to their chronic condition and do not exercise beyond their limits. Exercise testing, critical evaluation of the patient's functional class, and assessment of ventricular function will help to determine the timing, risk, and success of the operation.

Transplantation is sometimes needed when the cardiac anatomy is not suitable for an operation or when ventricular dysfunction is too severe. The indications for transplantation are similar to those in patients with other cardiac conditions, and should be considered in patients who have New York Heart Association functional class IV symptoms, despite optimal medical therapy and in the absence of other therapeutic options. The number of adults with CHD requiring heart transplantation is currently relatively small, and an even smaller group has been reported with heart and

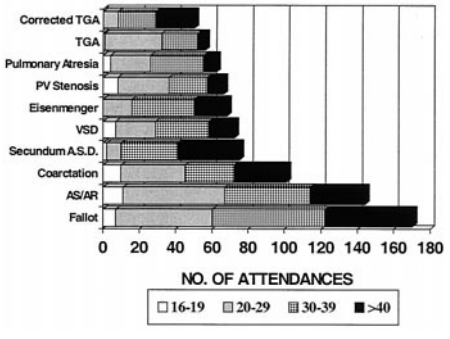

Figure 2. Outpatient attendance for 1997, according to age >16 years, basic diagnosis, and age. AR = aortic regurgitation; AS = aortic stenosis; ASD = atrial septal defect; PV = pulmonary valve; TGA = transposition of the great arteries; VSD = ventricular septal defect. Statistics from Jane Somerville, London, U.K.

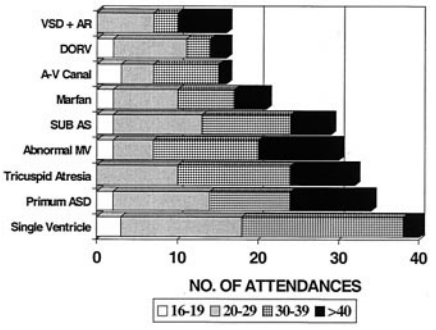

Figure 3. Outpatient attendance for 1997, according to age >16 years, basic diagnosis, and age. AR = aortic regurgitation; ASD = atrial septal defect; A-V = atrioventricular; DORV = double-outlet right ventricle; MV = mitral valve; SUB AS = subaortic stenosis; VSD = ventricular septal defect. Statistics from Jane Somerville, London, U.K.

lung transplantation. Transplantation in adults with CHD has been most frequently performed in patients with Fontan-type repair, transposition of the great arteries after a Mustard or Senning procedure with severe systemic (morphologically right) ventricular dysfunction, congenitally corrected transposition with ventricular dysfunction, severe Ebstein's anomaly or Eisenmenger syndrome. Transplantation needs may also increase in the next two decades, as more children with complex single-ventricle physiology undergo Fontan-like repair.

Electrophysiology. There is a growing recognition that arrhythmias, both atrial and ventricular, are an increasing problem in terms of management in these patients, and they are often associated with increasing morbidity and mortality. This is a consequence of: 1) underlying anatomic abnormalities; 2) chamber dilation and progressive fibrosis; 3) previous surgical incisions; and 4) compromised hemodynamic status. Pharmacologic management options for these patients may be limited by concomitant sinus node dysfunction, significant associated systemic ventricular dysfunction, and the desire for pregnancy.

Over last few years, newer, nonpharmacologic management options have emerged, specifically: 1) catheter ablation; 2) surgical approaches targeting structural abnormalities as well as offering intraoperative electrophysiologic ablation; and 3) automatic implantable internal defibrillators and a new generation of pacemakers with algorithms designed to prevent and treat atrial tachyarrhythmias. With some exceptions, in this population catheter ablation has met with only modest success so far; it is anticipated that ongoing refinements of mapping and ablation techniques will result in improved outcomes. A combined surgical approach has been employed successfully in the management of atrial arrhythmias, including those in patients with Ebstein's anomaly and patients undergoing Fontan revision, including the arrhythmias (both atrial and ventricular) seen after tetralogy of Fallot repair.

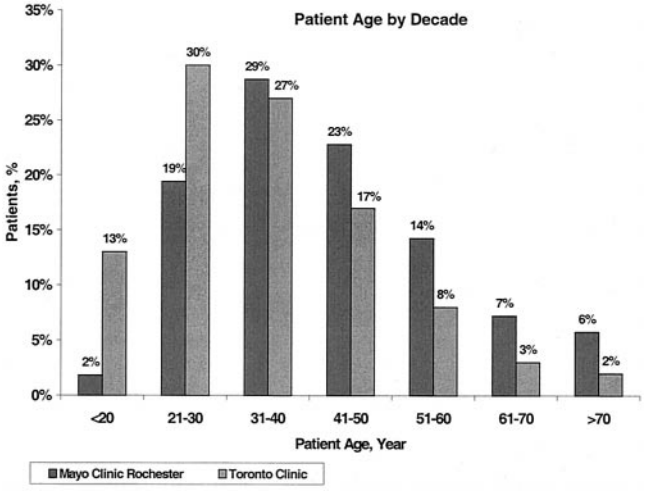

Figure 4. Age range of patients with CHD in two tertiary care centers.

Table 8. Cardiac Surgery for Congenital Heart Disease* by Major Diagnosis†

	Age ≥16 Years (n)
Atrial septal defect	131
Bicuspid aortic valves	129
Other diagnoses	28
Pulmonary atresia	23
Tetralogy of Fallot	20
Ebstein's anomaly	11
Transposition of the great arteries	10
Atrioventricular canal	9
Coarctation of the aorta	9
Double-outlet right ventricle	5
Anomalous pulmonary venous connection	4
Double-inlet left ventricle	4
Patent ductus arteriosus	3
Tricuspid atresia	2
Ventricular septal defect	2
Truncus arteriosus	1
Total	391

*Performed at the Mayo Clinic, Rochester, Minnesota, in 1999. †Includes all congenital diagnoses and all bicuspid aortic valve repairs.
The patients' mean age was 36.1 years (oldest patient 86 years).

These approaches, again, emphasize the desirability of a closely integrated collaboration between the surgeon, electrophysiologist, and cardiologist. With refinements in medical and nonpharmacologic therapy, it is anticipated that the need for arrhythmia therapy will increase in this aging population. The newer generation of atrial antitachycardia pacemakers and/or defibrillators will hopefully offer an expanded range of therapeutic options for these patients. However, issues of venous access, intracardiac shunts, and thromboembolic risk will often preclude a transvenous approach for lead implantation, and an epicardial approach may need to be considered. Data from current automatic implantable cardioverter-defibrillator trials in patients with ischemic or dilated cardiomyopathy appear to support expanded indications for automatic implantable cardioverter-defibrillator use in patients with substantial ventricular dysfunction, nonsustained ventricular tachycardia, and inducible ventricular tachycardia according to the electrophysiologic study. It is possible that these results may be extrapolated to adults with CHD, suggesting that the rate of

automatic cardioverter-defibrillator implantation will continue to increase in this patient population.

Catheterization/Intervention. Cardiac catheterization has been the diagnostic "gold" standard for CHD for the past 50 years. For the past 20 years, it has been increasingly supplemented by noninvasive diagnostic modalities; initially, cardiac ultrasound and, more recently, computed tomographic scanning and magnetic resonance imaging. Advances in these technologies have been logarithmic, and it is likely that in the coming decade, both morphologic and functional assessments of this patient population will be increasingly accomplished noninvasively.

Today, diagnostic catheterization is largely reserved for resolution of specific issues concerning operative interventions, including: 1) the preoperative evaluation of coronary arteries; 2) the assessment of pulmonary vascular disease and its response to vasoactive agents for planned, traditional surgical intervention and/or heart or heart/lung transplantation; and 3) as an adjunct to the noninvasive assessment of the morphologic and functional characteristics of many complex congenital lesions (e.g., delineation of arterial and venous anatomy, patients with heterotaxy, Fontan candidates, and patients who have had previous palliation in the form of a shunt). Such procedures should be performed by experienced and trained operators who maintain an adequate minimal volume annually.

Evaluation for possible interventional catheterization has become an increasingly common indication for diagnostic catheterization. For some lesions, notably valvular pulmonary stenosis, branch pulmonary stenosis, residual or recurrent aortic coarctation, and arteriovenous fistulae, catheter intervention is widely considered to be the treatment of choice. Coil or device occlusion of the patent ductus produces results comparable to those of surgical closure, and device closure of secundum atrial septal defects is often employed, although the success rate varies with operator expertise and the specific device used. It is likely that technical problems related to these devices will ultimately be overcome. Dilation of stenotic palliative shunts can obviate the need for re-operation, and transcatheter occlusion of shunts before repair of intracardiac lesions may simplify the surgical procedure. Along with the growth of interventional catheterization, there has been a renewed interest in small-incision cardiac surgery, and there will likely be continued advocacy for both management alternatives. Finally, a national and global perspective must be kept in mind, relative to limited resources in developing regions where interventional catheterization may provide partial or definitive treatment for many patients with CHD who do not have access to cardiac surgery.

Echocardiography. With improvements and refinements in echocardiographic technology, most adults attending an outpatient clinic undergo transthoracic echocardiography and, when necessary, complementary transesophageal echocardiography and magnetic resonance imaging. Two-dimensional imaging is more challenging in this patient population because of larger body size and often multiple previous surgical scars. The use of transesophageal echocardiography intraoperatively is also increasing, and it has been shown that it has a major impact on cardiac surgical procedures in 6% to 9% of cases (i.e., that it is desirable or necessary for the patient to resume cardiopulmonary bypass for revision of the cardiac procedure). Physicians interpreting these echocardiograms need to be experienced and have expertise in all aspects of CHD.

A high rate of diagnostic errors in pediatric echocardiograms performed in community-based adult laboratories has been reported (8). This study reported patients of varying ages, from one day to 18 years, and either interpretive or technical errors that were of major or moderate importance occurred in 53% of cases. There is reason to believe that in older patients, errors occur even more frequently because image acquisition is more challenging. Clearly, both expertise and technology are necessary to provide the best care.

Conclusions. The data, estimates, and models described herein emphasize that patients in the U.S. have been underserved by the present health care system. Over the next decade, a more comprehensive system must be developed for this growing population, with considerable collaboration between cardiologists specializing in pediatrics and adults. This Conference will facilitate the further recognition of these needs and hopefully help to develop the resources needed to achieve these objectives.

TASK FORCE 1 REFERENCE LIST

1. Moller JH, Taubert KA, Allen HD, Clark EB, Lauer RM, a Special Writing Group from the Task Force on Children and Youth, American Heart Association. Cardiovascular health and disease in children: current status. Circulation 1994;89:923–30.
2. Wren C, O'Sullivan JJ. Future demand for follow-up of adult survivors of congenital heart disease. Heart 2001;85:438–43.
3. Hoffman JI, Christianson R. Congenital heart disease in a cohort of 19,502 births with long-term follow-up. Am J Cardiol 1978; 42:641–7.
4. Fyler DC. Report of the New England Regional Infant Cardiac Program. Pediatrics 1980;65 Pt 2:375–461.
5. Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995;70:50–4.
6. Stark J. Glenn Lecture: how to choose a cardiac surgeon. Circulation 1996;94 Suppl II:II-1–4.
7. Dore A, Glancy DL, Stone S, Menashe VD, Somerville J. Cardiac surgery for grown-up congenital heart patients: survey of 307 consecutive operations from 1991 to 1994. Am J Cardiol 1997;80:906–13.
8. Stanger P, Silverman NH, Foster E. Diagnostic accuracy of pediatric echocardiograms performed in adult laboratories. Am J Cardiol 1999; 83:908–14.

JACC Vol. 37, No. 5, 2001
April 2001:1161–98

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

Elyse Foster, MD, FACC, *Co-Chair,* Thomas P. Graham, Jr., MD, FACC, *Co-Chair*, David J. Driscoll, MD, FACC, Graham J. Reid, PHD, CPsych, John G. Reiss, PHD, Isobel A. Russell, MD, PHD, Matthew Sermer, MD, Samuel C. Siu, MD, SM, FACC, Karen Uzark, PHD, PNP, Roberta G. Williams, MD, FACC, Gary D. Webb, MD, FACC

MANAGING THE TRANSITION TO ADULTHOOD BEGINS IN CHILDHOOD

Transition into the adult health care system is crucial for patients with congenital heart disease (CHD), as well as for adolescents with many other chronic conditions. Indeed, "arranging efficient and caring transfer for adolescents from pediatric to adult care (is). . .one of the great challenges facing pediatrics—and indeed the health care services—in the coming century" (1). Centers that care for adolescents and young adults with CHD need to develop structured plans for the transfer of care from the department of pediatric cardiology to that of adult cardiology. A comprehensive program taking a developmental approach beginning in childhood and adolescence should achieve better results than programs that focus only on the transfer to adult care at a specified age.

Currently, sufficient empirical data are not available to support the identification of "best practices" regarding transition in this patient group. However, descriptive and qualitative studies have indicated that the key elements of an effective transition program include:

1. A policy on timing of transfer to adult care (age 18 or upon leaving school is recommended by many, with some flexibility);
2. A preparation period and education program that focus on a set of skills that enables young people and their families to function in an adult clinic (e.g., understanding the disease, treatment rationale, and source of symptoms; recognizing deterioration and taking appropriate action; learning how to seek help from health professionals and how to operate within the medical system);
3. A coordinated transfer process (including a detailed written plan and pretransfer visit to the adult clinic, with an introduction to the adult provider and with a designated coordinator such as a clinic nurse);
4. An interested and capable ACHD regional center that is at least equivalent in quality to that of the pediatric source the patient is leaving (see subsequent discussion);
5. Administrative support; and
6. Primary care involvement.

The published data also suggest that transitions are more successful in health care settings where:

1. The preparation for transition begins before adolescence, and transition is seen as an essential component of high-quality health care.
2. There is a formal transition program.
3. Young people are not transferred to adult services until they have the necessary skills to function in an adult service and have finished growth and puberty.
4. There is an identified person on both the pediatric and adult teams who has responsibility for transition arrangements (usually nurse specialists).
5. Management links are developed between the pediatric and adult systems, and financial and contracting issues are worked out in detail and put in writing.
6. The evaluation of transition arrangements is undertaken as part of a continuous quality improvement process.
7. Transfer is planned and carried out during a period of medical stability; and
8. After transfer, there should be ongoing consultation with the referring pediatric cardiologist.

Sometimes parents need counseling and support to let go of their adult offspring. Adolescents need support and encouragement to begin making decisions, maximizing their strengths and abilities, and taking control of their lives. Support groups for patients and/or parents may provide a reality-based interchange of shared issues. Large referral institutions can provide direct support group access, but patients in other areas may be best served by Internet support groups (e.g., CACHnet [www.cachnet.org], Children's Health Information Network [http://tchin.org]) and newsletters.

Factors such as a lack of symptoms and strong attachment to a pediatric cardiology program may result in the failure to transfer successfully to ACHD services. Many adults with CHD have the impression that the operation performed in childhood was a "cure." They may be unaware of their prognosis and believe that regular cardiac follow-up is unnecessary. Comprehensive, individualized education regarding their cardiac condition and health care needs, as well as a "road map" regarding life-style implications, can help patients to achieve independence. In addition to routine health care recommendations, the health education objectives should be specific for young adults with CHD and should include specific information on diagnoses and operations, medications and their side effects, endocarditis prophylaxis, exercise prescription, contraception and family

planning, career planning and resources, insurance, guidelines for frequency of medical follow-up, and dental care. Education regarding symptoms that could be serious, such as arrhythmias, is essential. This information should be summarized in a "health passport" that can be held in the patient's possession.

The timing and manner of communicating with patients is very important. Presenting an overwhelming amount of information in a single session should be avoided, especially during early transition visits. The potential to throw the patient into despair or denial exists and could lead to avoidance of much-needed visits to a medical center in the future. Discussions should proceed at a rate commensurate with the psychosocial development and circumstances of the patient. Traditionally, many pediatricians have continued to care for adults with developmental disabilities, but provision for this group needs to be part of the planning for adult care.

Children with CHD should be given the expectation that they will grow up to be healthy and able to work. Questions like "what do you want to be when you grow up?" should be asked by pediatric primary and specialty care providers, starting in early childhood (~age 4). In their transition to adulthood, adolescents should prepare for economic independence and vocational competence. Although the majority of patients with postoperative CHD are functionally normal, the label of heart disease or a mild disability can heighten these developmental challenges. The possibility of further surgery can also impede long-term planning. Adolescents should be encouraged to achieve higher education and skills necessary for employment in occupations that are reasonable in relation to their work capacity. Vocational guidance should be emphasized in *early* adolescence (ages 13–15), as training and acquisition of special knowledge and skills are important to the young adult if he or she is going to compete with other applicants.

UNIQUE MEDICAL ISSUES

Cyanotic patients. Cyanotic patients with CHD should be seen regularly at a regional ACHD center. Their physicians should be aware of special issues in their care. There are medical problems extrinsic to the cardiovascular system, which can cause significant morbidity and mortality in such patients. Severe cyanosis leads to marked erythrocytosis and, frequently, to low platelet counts (>100,000) (2), which, fortunately, seldom lead to significant bleeding. The absence of erythrocytosis (e.g., hemoglobin >17.0 g/dl) in such patients should raise concern about a "relative anemia" and its cause and implications. Excessive erythrocytosis adversely affects whole blood viscosity, but this problem is normally not associated with symptoms until the hematocrit levels are at least >60% to 65%. The only valid indication for therapeutic phlebotomy is to alleviate symptoms of hyperviscosity. Many patients undergo phlebotomy by primary care physicians to reduce their hematocrit. This practice should be discouraged; phlebotomy should be undertaken only in a center that manages cyanotic patients. When blood is removed, volume replacement with normal saline is recommended. Failure to follow this procedure can be associated with hypotension, increased right to left shunting, stroke, seizures, and death, especially in patients with pulmonary vascular disease. Multiple phlebotomies result in iron depletion, which is disadvantageous and has an independent negative effect on exercise performance. Iron deficiency is associated with impaired small-vessel blood flow and an increase in the risk of reversible ischemic neurologic deficits and stroke (3). When iron deficiency does occur, it should be treated.

Cyanotic patients frequently have hyperuricemia with arthralgia, gouty arthritis, and overt tophaceous deposits of urate. These abnormalities are secondary to low uric acid fractional excretion, not to urate overproduction (4). Therefore, hyperuricemia is a marker of abnormal renal function, which can be a problem in patients with long-standing cyanotic heart disease. Acute gouty arthritis responds to colchicine; special care should be taken to avoid dehydration that could occur with vomiting and diarrhea, which would require cessation of drug therapy. Allopurinol, with or without a uricosuric agent, is considered for symptomatic hyperuricemia refractory to other medications (2).

Pulmonary vasodilator therapy, specifically prostacyclin, has been used successfully in a small number of patients with Eisenmenger syndrome, serving as a bridge to repair in a few. The role for pulmonary vasodilators is being studied intensively.

Noncardiac surgery. Adults with moderate and complex CHD (see Tables 4 and 5 of Task Force #1) who require noncardiac surgery have special needs to be addressed by the surgical and anesthesia team. Ideally, operations in patients with complex CHD should be performed at a regional ACHD center with physicians experienced in the care of these individuals and with the consultation of cardiologists trained in this discipline (5). Frequently encountered management issues include cessation of anticoagulant agents and use of antibiotics for endocarditis prophylaxis (6). Important considerations for anesthetic management include the functional class of the patient, ventricular function, persistent shunts, valvular disease, arrhythmias, erythrocytosis, pulmonary disease, and pulmonary vascular disease.

Risk factors that help predict the possibility of perioperative risk include cyanosis ($p = 0.002$), treatment for congestive heart failure ($p < 0.001$), poor general health ($p < 0.001$), and younger age ($p = 0.03$) (7). Patients with pulmonary hypertension probably have a higher complication rate (15%) than patients without pulmonary hypertension (4.7%; $p = 0.08$). Procedures performed on the respiratory and nervous systems seem to be associated with the most complications.

The extent of preoperative evaluation varies depending on the complexity of the heart disease. A complete understanding of the patient's underlying anatomy is necessary. A preoperative echocardiogram and, rarely, cardiac catheter-

ization may be indicated when recent data are not available. Stress studies may be indicated to exclude coexistent coronary artery disease in older adults. Estimates of pulmonary hypertension are useful, because some patients are at increased risk for Eisenmenger syndrome (8). If pulmonary disease exists, preoperative pulmonary function tests may be necessary to determine its severity and to estimate the need for postoperative ventilation. Preoperative laboratory testing in cyanotic patients should include an evaluation of the hematologic system (5), including coagulation and platelet abnormalities (9). Isovolumetric phlebotomy to a hematocrit <65% has been recommended to improve hemostasis. Practices vary among centers, and further study is warranted.

In patients receiving long-term anticoagulation, protocols for stopping warfarin, by using perioperative heparin, and restarting warfarin should be developed and coordinated with the surgical and dental team to minimize blood loss and prevent complications. The decision for invasive monitoring, such as intra-arterial catheters and/or central venous catheters, should be based on the magnitude of the operation and the specific nature of the cardiac defect. The decision to monitor invasively should be weighed against the risk of complications. In all cyanotic patients, meticulous attention should be paid to all intravenous lines to ensure freedom from air bubbles, which may cause systemic air embolism. Intraoperative transesophageal echocardiography may be useful for continuous monitoring of ventricular function and for estimating preload conditions. The choice of anesthetic agent depends on the severity of the cardiac disease and other co-morbidities and must be tailored to the operation. Avoidance of myocardial depression and hypovolemia is emphasized. Epidural anesthesia can provide excellent operative and postoperative analgesia, with minimal cardiovascular side effects in select patients.

CONTRACEPTION AND PREGNANCY IN WOMEN

Contraception. All patients must be well informed of the risks of pregnancy associated with their condition and the available options to avoid pregnancy when desired. The risks of pregnancy vary widely among the specific types of CHD (see subsequent discussion). There are no systematic outcome data on the safety of contraceptive methods in women with CHD. The choice of contraceptive method is usually made by the patient. However, it is the responsibility of the physician to provide thorough counseling about the risk of unplanned pregnancy in the case of non-compliance, poor acceptance or failure of the contraceptive technique, and any risk associated with the specific method including infective and thromboembolic complications. In considering surgical sterilization because of high risk, the patient should be fully informed of the potential for medical advances that may permit future pregnancy at lower risk.

Pregnancy risk. Pregnancy in women with CHD not complicated by Eisenmenger syndrome is associated with a low mortality (10–12). However, potential risk factors for maternal morbidity include poor maternal functional class, poorly controlled arrhythmias, heart failure, cyanosis, significant left heart obstruction, and a history of cerebral ischemia (10–14). Cyanosis is a risk factor for fetal and neonatal complications (10–12,14). On the basis of these risk factors, patients can be stratified into low-, intermediate-, or high-risk categories (12). An absence of these risk factors would generally place patients into a low-risk category. The highest risk is associated with Eisenmenger syndrome, in which postnatal maternal mortality can exceed 50%. Because much of the current data are based on retrospective case series from tertiary care institutions, one should exercise caution in risk stratification of pregnant women with uncommon conditions such as Mustard/Senning or in those who have had a Fontan procedure (15–17). Patients with these lesions or procedures should be placed in the intermediate-risk category until additional data become available. The risk of in vitro fertilization for surrogate pregnancy in high-risk women with CHD has not been defined. Medical or surgical termination of pregnancy in intermediate or high-risk patients requires careful monitoring, and preferably it should be done in a regional ACHD center.

Special needs of pregnancy. Women with heart disease who are at intermediate or high risk or an uncertain level of risk for complications should be managed in a high-risk perinatal unit by a multidisciplinary team including an obstetrician, cardiologist, anesthesiologist, and pediatrician. The team should meet early in the patient's pregnancy to review the cardiac lesion, anticipated effects of pregnancy, and potential problems and to develop a management plan. Specific issues that need to be considered include the timing and mode of delivery, the type of anesthesia to be used, the need for hemodynamic monitoring before and after delivery and the use of antibiotic prophylaxis. Women with heart disease in the low-risk group can usually be managed in a community hospital setting.

Risk of recurrence of CHD in offspring. Genetic counseling regarding etiology, inheritance, recurrence risk, and prenatal diagnosis options should be made available to all patients with CHD. It is important to obtain the patient's prenatal and postnatal history, including maternal exposure to teratogens, as well as a detailed family history, and to perform a thorough examination looking for congenital abnormalities (18).

In all women contemplating pregnancy, exposure to teratogens should be investigated; in some cases, finding an alternative medication will be necessary. Angiotensin-converting enzyme inhibitors and angiotensin II receptor antagonists should not be used during pregnancy. Medications for which substitution should be considered include warfarin and amiodarone. No medications, including over-the-counter preparations, should be taken during pregnancy without physician approval. Preconception consumption of

multivitamins including folic acid decreases the incidence of CHD (19).

Knowledge of the genetic basis of CHD is expanding rapidly. The role of genetic testing is evolving, and genetic counseling should be made available. The recurrence rate of CHD in offspring is variable, ranging from 3% to 50%. A higher recurrence risk when the mother rather than the father is affected has raised the possibility of mitochondrial inheritance in some patients (20). Diseases with a single gene disorder and/or chromosomal abnormalities are associated with a high recurrence rate. In Marfan, Noonan, and Holt-Oram syndromes, there is a 50% risk of recurrence.

Fetal echocardiography at 16 to 18 weeks gestation should be available to all patients with CHD. Chorionic villus sampling or amniocentesis may be useful after discussion of the potential risks and benefits.

EXERCISE TOLERANCE AND REHABILITATION

Exercise data. The ability to exercise is one measure of quality of life, and it is used to assess the effect of disease, the results of treatment, and the ability to tolerate the stress associated with pregnancy or needed surgery. There have been numerous studies of exercise tolerance in children and adolescents with CHD but very few studies in adults.

Adults with pulmonary stenosis have well-preserved but still subnormal exercise tolerance. Exercise tolerance for adults with aortic stenosis or a ventricular septal defect (VSD) is subnormal and even less than that for adults with pulmonary stenosis. Rather surprisingly, two investigators have reported that exercise tolerance is subnormal for patients who had repair of an atrial septal defect (ASD). Reybrouck et al. (21) have demonstrated that the age when closure of an ASD is performed influences postoperative exercise tolerance. Adults with complex conditions, such as Ebstein's anomaly or a single ventricle, have a markedly abnormal exercise tolerance. There are few studies of exercise tolerance of adults with transposition of the great arteries, pulmonary atresia with or without VSD, and other complex conditions. However, studies of children and adolescents with these defects show subnormal exercise tolerance, and it is assumed that exercise tolerance would be no greater in adults with similar defects.

Recommendations for athletic participation for patients with CHD were published in the 26th Bethesda Conference (22) and are the best consensus data available.

Exercise training and rehabilitation. Because adults with CHD have subnormal exercise tolerance, an obvious question is whether physical conditioning reduces symptoms and improves exercise tolerance and quality and/or length of life. These issues have not been studied. There are numerous studies validating the benefits of exercise for healthy adults and those with coronary artery disease (23). There have been several studies of the utility of exercise programs for children with CHD (24,25). Despite major design problems, these studies demonstrate that a structured rehabilitation program can increase exercise efficiency. Improved exercise performance (i.e., maximal oxygen consumption) was not demonstrated in most studies. Because all of these studies were small, a survival benefit could not be demonstrated. An alternative to a costly structured rehabilitation program is a simple home program of physical rehabilitation, which was successful in one study (26). The efficacy and safety of a structured exercise rehabilitation program for adults with CHD are unknown. Issues that require further study in adult patients include the efficacy of such a program in improving fitness and aerobic capacity, the safety of such programs, and the interaction between congenital and acquired heart disease.

PSYCHOSOCIAL ISSUES

Only recently have patients with complex CHD survived into adult life in large numbers. Their survival creates hope that continuing advances will help them maintain both quality of life and longevity. However, patients may experience despair due to their awareness of residual morbidities and the knowledge of possible or probable early mortality, or limitations in their social lives and educational or occupational attainment. Healthy psychosocial functioning depends on their ability to balance hope and despair. Adults with CHD must also confront both CHD-specific and general developmental tasks. Psychosocial issues may be affected by lesion severity (simple vs. complex), visibility (e.g., cyanosis), and functional disability (27).

Life-span development and CHD. Table 1 is a proposed model outlining developmental tasks faced by individuals with CHD, beginning in adolescence; this model could be tested in future studies.

Physical development. Adults with CHD may struggle with physical appearance (e.g., scars, smaller body size, cyanosis, clubbing), physical limitations, and acute or gradual decreases in physical functioning (28–30). Physical decline may be difficult to deal with, as peers are often less able to empathize with these changes.

Social and family relations. Adults with CHD are less likely to be married or cohabiting or to have children and are more likely to live with their parents, as compared with healthy peers (31,32). These differences may reflect life-style decisions made based on beliefs or knowledge regarding shortened life-expectancy, concerns about pregnancy risks, economic constraints, or the need for social support (33,34). Patients limiting themselves due to misinformation need counseling. Difficulty discussing CHD issues with family or friends is common among adolescents and young adults with CHD, especially among patients whose parents rarely discussed their own disease (35). Patients and their families may need assistance in finding a balance between independence and interdependence that optimizes the psychosocial and physical health of the patient with CHD. Adolescent and young adult patients, in particular, may need assistance

Table 1. Life-Span Developmental Tasks and Issues for Adolescents and Adults With Congenital Heart Disease

	Developmental Tasks by Age Group			
Domains	**Mid-Adolescence (14–16 years)**	**Late Adolescence (16–19 years)**	**Young Adulthood (19–35 years)**	**Mid-Adulthood (mid 30s+)***
Physical	Coping with body image and limitations in physical functioning		Gradual or abrupt decreases in physical functioning; burden/complications with onset of common illnesses of adulthood	
Social and family relations	Peer acceptance of physical appearance/limitations; coping with stigmatization; lack of social support for CHD issues	Decisions about dating; increasing independence from family; lack of social support for CHD issues	Decisions regarding life partner and reproduction; coping with loss of normative family life cycle; finding a social group/network	Addressing the impact of premature death of partner, any children, and extended family
Emotional	Managing anxiety-provoking medical procedures; maintaining emotional adjustment during period of critical transitions		Managing anxiety-provoking medical procedures; avoiding arrhythmia-related anxiety/phobic reactions; avoiding despair, depression, or anxiety; maintaining emotional/mental health	
Education and vocation	Coping with possible intellectual and/or learning disabilities	Selecting educational and vocational goals appropriate to present/future abilities	Stigmatization/discrimination in obtaining employment; maintaining employment during medical crises	Maintaining/changing employment and/or career goals with decreases in physical functioning
Medical	Taking some responsibility for medical care; learning appropriate health behaviors	Increasing responsibility for medical care; transition to adult care; knowledge of diagnosis, prognosis, and associated health behaviors	Primary responsibility for medical care; knowledge of prognosis; re-operation(s); CHD complications; coping with medical procedures and hospitalization; coping with procedure-related pain	
Health behaviors	Avoiding initiation of risky health behaviors; maintaining appropriate weight and getting exercise; maintaining oral hygiene and preventing endocarditis	Regular medical follow-up; avoiding risky health behaviors; maintaining appropriate weight; getting appropriate exercise; maintaining oral hygiene and preventing endocarditis		
Personality and identity	Integrating CHD into self; accepting being different and unique	Lack of control over health outcomes; increasing independence	Balancing independence and interdependence with family and friends	Resolving loss of typical life achievements; facing prospect of premature death

*Life-expectancy varies with lesion severity and is increasing with improved medical care.

in ways to educate their peers regarding CHD and to maintain a peer network.

Emotional health. Although most adolescents and adults with CHD appear to be free of psychopathology, there are too few studies to draw conclusions about the emotional health of this group of patients. Results of comparisons of emotional adjustment between patients with CHD and healthy peers have been mixed. One study found that when items likely reflecting CHD symptoms (e.g., dizziness) were removed from analyses, group differences were no longer significant (36).

In patients with acquired heart disease, depression and anxiety are linked to an increased risk of cardiac and all-cause mortality and sudden cardiac death (37–40). Given the high prevalence of arrhythmias with complex CHD, this potential relationship should be examined. Twenty percent of all adults have a psychiatric disorder (41). Even if adults with CHD were not at increased risk of psychopathologic conditions, one in five patients would be expected to have a significant mental health problem. Surgery, hospital admission, invasive medical procedures, and even routine appointments may trigger emotional distress, especially in individuals with a pre-existing emotional disorder.

Medical issues. Only one half to three fourths of adults with CHD can correctly state or describe their diagnosis (42–44). Given complex anatomies and surgical repairs, it may not be possible for all patients with CHD to have precise anatomic knowledge, but aids such as a "health passport" may be helpful in providing the patient with the most important information.

Adults with CHD may have difficulty coping with repeated hospital admissions, operations, and other painful medical procedures. They underwent cardiac surgery during an era of inadequate pain control (45), which may result in centrally mediated pain sensitization for them (46). Patients with CHD should receive education about pain and its management and receive optimal medication and management of procedure-related pain. Health care personnel must also be informed of the special issues related to pain management in this group.

Health behaviors. Poor knowledge of behaviors related to endocarditis and its prevention are common (42–44,47). Unrealistic fears (e.g., fear of damaging the heart or having a cardiac arrest) may be a factor in their lack of activity. However, some patients who have been advised against heavy exercise will engage in it anyway (e.g., by engaging in contact sports) (30). Although consensus (27) and common sense suggest that regular medical follow-up, as compared with nonattendance, should result in better outcomes, proof is lacking. Patients' beliefs regarding treatment effectiveness (e.g., medication) or health behaviors (e.g., regular medical appointments) are a significant predictor of compliance and should be assessed.

Personality and identity. Establishing one's identity, balancing independence with interdependence, and accepting death are tasks of normal development. In addition, adults with CHD must incorporate their condition into their identity, deal with a lack of control over changes in physical functioning, resolve the loss or disruption of typical developmental achievements (e.g., surgery may result in loss of an academic year), and face the prospect of premature mortality. These issues must be faced repeatedly throughout adulthood and may necessitate counseling.

Screening and prevention. Routine screening for psychosocial or physical problems is not without risk (48–50) and should be undertaken only if there are accurate measures, appropriate mechanisms to provide feedback, and appropriate resources for treatment. Because validated CHD-specific measures do not exist, measures of perceived risk of CHD complications and health behaviors should be developed.

Treatment issues. Although there are validated psychosocial and pharmacologic treatments for many psychological disorders (41), treatment effectiveness could be enhanced if interventions are adapted to deal with CHD-specific issues. Treatments specific to CHD should focus on enhancing knowledge, modifying maladaptive beliefs, and dealing with periods of transition and acute stress.

The level of treatment intensity could be tailored to the severity of the problem, ranging from self-help materials for those with mild or moderate problems to individual or group therapy for those with severe problems. Creative solutions that offer individual counseling for patients at a distance from a regional ACHD center are needed. As interventions are developed, it is crucial to document procedures and evaluate effectiveness so centers can share and build on each other's experiences.

RECOMMENDATIONS

Transition to Adult Life

- Structured plans should be developed to transition patients from pediatric to adult CHD care. Transition to a regional ACHD center can be difficult for patients, and the presentation of an overwhelming amount of information in a single session should be avoided. Discussions should proceed at a rate commensurate with the psychosocial development and circumstances of the patient.
- Individual patient education regarding his/her diagnosis and specific health behaviors should be a priority.
- Important historic information, including comprehensive diagnostic data, procedures, operations, and medications, should be kept by the patient as a summary of past and present important health issues. The American College of Cardiology should support development of a health care "passport," which would be useful for all patients with CHD and their health care providers.
- A continuum of vocational assessment beginning in childhood should be available for patients with CHD and should be continued during the developmental, adolescent, and young adult years.

Noncardiac Surgery

- Ideally, noncardiac operations on patients with complex CHD should be performed at a regional ACHD center with the consultation of an anesthesiologist with experience in CHD, particularly for more complex surgery or for patients with adverse risk factors that include poor functional class, pulmonary hypertension, CHF, and cyanosis.

Reproductive Issues

- Contraceptive counseling must be available, when appropriate, to all patients with CHD.
- A multidisciplinary team at a regional ACHD center is needed for pregnancy and delivery, as well as for the management of indeterminate-, intermediate-, or high-risk patients.
- The recurrence risk of CHD is highly variable and should be discussed prospectively with all patients. Genetic counseling should be made available through regional ACHD centers.

Exercise and Rehabilitation

- Guidance for athletic participation for patients with CHD should be in accordance with the published recommendations of the 26th Bethesda Conference report, which represents the best consensus data available.
- The efficacy and safety of exercise rehabilitation programs in adults with CHD have not been studied, and research in this area should be supported.

Psychosocial Issues

- The emotional health of adults with CHD should be a priority in the overall care of this patient population. Appropriate screening and referral sources for treatment should be available at all regional ACHD centers.
- Tools for screening of psychosocial problems in this population should be developed and tested.
- Data should be developed to assess the impact of regular follow-up care on the long-term physical and psychosocial health of adults with CHD.
- Available professionals and facilities for the treatment of psychological disorders are scarce, and creative solutions for counseling patients in groups and/or those at a distance from home should be developed.

TASK FORCE 2 REFERENCE LIST

1. Viner R. Transition from pediatric to adult care: bridging the gaps or passing the buck? (see comments) Arch Dis Child 1999;81:271–5.
2. Perloff JK, Rosove MH, Child JS, Wright GB. Adults with cyanotic congenital heart disease: hematologic management. Ann Intern Med 1988;109:406–13.
3. Ammash N, Warnes CA. Cerebrovascular events in adult patients with cyanotic congenital heart disease. J Am Coll Cardiol 1996;28: 768–72.
4. Ross EA, Perloff JK, Danovitch GM, Child JS, Canobbio MM. Renal function and urate metabolism in late survivors with cyanotic congenital heart disease. Circulation 1986;73:396–400.
5. Baum VC, Perloff JK. Anesthetic implications of adults with congenital heart disease. Anesth Analg 1993;76:1342–58.
6. Dajani AS, Taubert KA, Wilson W, et al. Prevention of bacterial endocarditis: recommendations by the American Heart Association. Circulation 1997;96:358–66.
7. Warner MA, Lunn RJ, O'Leary PW, Schroeder DR, the Mayo Perioperative Outcomes Group. Outcomes of noncardiac surgical procedures in children and adults with congenital heart disease. Mayo Clin Proc 1998;73:728–34.
8. Ammash NM, Connolly HM, Abel MD, Warnes CA. Noncardiac surgery in Eisenmenger syndrome. J Am Coll Cardiol 1999;33:222–7.
9. Colon-Otero G, Gilchrist GS, Holcomb GR, Ilstrup DM, Bowie EJ. Preoperative evaluation of hemostasis in patients with congenital heart disease. Mayo Clin Proc 1987;62:379–85.
10. Whittemore R, Hobbins JC, Engle MA. Pregnancy and its outcome in women with and without surgical treatment of congenital heart disease. Am J Cardiol 1982;50:641–51.
11. Shime J, Mocarski EJ, Hastings D, Webb GD, McLaughlin PR. Congenital heart disease in pregnancy: short- and long-term implications. Am J Obstet Gynecol 1987;156:313–22.
12. Siu SC, Sermer M, Harrison DA, et al. Risk and predictors for pregnancy-related complications in women with heart disease. Circulation 1997;96:2789–94.
13. Lao TT, Sermer M, MaGee L, Farine D, Colman JM. Congenital aortic stenosis and pregnancy: a reappraisal. Am J Obstet Gynecol 1993;169:540–5.
14. Presbitero P, Somerville J, Stone S, Aruta E, Spiegelhalter D, Rabajoli F. Pregnancy in cyanotic congenital heart disease. Outcome of mother and fetus. Circulation 1994;89:2673 (Published erratum appears in Am J Obstet Gynecol 1987;156:1361.)
15. Clarkson PM, Wilson NJ, Neutze JM, North RA, Calder AL, Barratt-Boyes BG. Outcome of pregnancy after the Mustard operation for transposition of the great arteries with intact ventricular septum. J Am Coll Cardiol 1994;24:190–3.
16. Genoni M, Jenni R, Hoerstrup SP, Vogt P, Turina M. Pregnancy after atrial repair for transposition of the great arteries. Heart 1999;81: 276–7.
17. Canobbio MM, Mair DD, van der Velde M, Koos BJ. Pregnancy outcomes after the Fontan repair. J Am Coll Cardiol 1996;28:763–7.
18. Siu S, Chitayat D, Webb G. Pregnancy in women with congenital heart defects: what are the risks? (editorial). Heart 1999;81:225–6.
19. Czeizel AE. Reduction of urinary tract and cardiovascular defects by periconceptional multivitamin supplementation. Am J Med Genet 1996;62:179–83.
20. Nora JJ. From generational studies to a multilevel genetic–environmental interaction. J Am Coll Cardiol 1994;23:1468–71.
21. Reybrouck T, Bisschop A, Dumoulin M, van der Hauwaert LG. Cardiorespiratory exercise capacity after surgical closure of atrial septal defect is influenced by the age at surgery. Am Heart J 1991;122: 1073–8.
22. Maron BJ, Isner JM, McKenna WJ. 26th Bethesda Conference: recommendations for determining eligibility for competition in athletes with cardiovascular abnormalities. Task Force 3: Hypertrophic cardiomyopathy, myocarditis and other myopericardial diseases and mitral valve prolapse. J Am Coll Cardiol 1994;24:880–5.
23. Fletcher GF, Balady G, Blair SN, et al. Statement on exercise: benefits and recommendations for physical activity programs for all Americans. A statement for health professionals by the Committee on Exercise and Cardiac Rehabilitation of the Council on Clinical Cardiology, American Heart Association. Circulation 1996;94:857–62.
24. Calzolari A, Drago F, Gagliardi G, et al. Rehabilitation of programs for children after congenital heart disease surgery. Cardiologia 1989; 34:587–92.
25. Driscoll D. Exercise rehabilitation programs for children with congenital heart disease: a note of caution. Pediatric Exercise Science 1990;2:191–6.
26. Longmuir PE, Turner JA, Rowe RD, Olley PM. Postoperative exercise rehabilitation benefits children with congenital heart disease. Clin Invest Med 1985;8:232–8.
27. Connelly MS, Webb GD, Somerville J, et al. Canadian Consensus Conference on Adult Congenital Heart Disease, 1996. Can J Cardiol 1998;14:395–452.
28. Tong EM, Sparacino PS, Messias DK, Foote D, Chesla CA, Gilliss

CL. Growing up with congenital heart disease: the dilemmas of adolescents and young adults. Cardiol Young 1998;8:303–9.
29. Gantt LT. Growing up heartsick: the experiences of young women with congenital heart disease. Health Care Women Int 1992;13: 241–8.
30. Schneider MB, Davis JG, Boxer RA, Fisher M, Friedman SB. Marfan syndrome in adolescents and young adults: psychosocial functioning and knowledge. J Dev Behav Pediatr 1990;11:122–7.
31. Utens EM, Verhulst FC, Erdman RA, et al. Psychosocial functioning of young adults after surgical correction for congenital heart disease in childhood: a follow-up study. J Psychosom Res 1994;38:745–58.
32. Kokkonen J, Paavilainen T. Social adaptation of young adults with congenital heart disease. Int J Cardiol 1992;36:23–9.
33. Van Tongerloo A, De Paepe A. Psychosocial adaptation in adolescents and young adults with Marfan syndrome: an exploratory study. J Med Genet 1998;35:405–9.
34. Alden B, Gilljam T, Gillberg C. Long-term psychological outcome of children after surgery for transposition of the great arteries. Acta Paediatr 1998;87:405–10.
35. Horner T, Liberthson R, Jellinek MS. Psychosocial profile of adults with complex congenital heart disease. Mayo Clin Proc 2000;75:31–6.
36. Casey FA, Sykes DH, Craig BG, Power R, Mulholland HC. Behavioral adjustment of children with surgically palliated complex congenital heart disease. J Pediatr Psychol 1996;21:335–52.
37. Denollet J, Brutsaert DL. Personality, disease severity, and the risk of long-term cardiac events in patients with a decreased ejection fraction after myocardial infarction. Circulation 1998;97:167–73.
38. Kawachi I, Colditz GA, Ascherio A, et al. Prospective study of phobic anxiety and risk of coronary heart disease in men. Circulation 1994; 89:1992–7.
39. Moser DK, Dracup K. Is anxiety early after myocardial infarction associated with subsequent ischemic and arrhythmic events? Psychosom Med 1996;58:395–401.
40. Frasure-Smith N, Lesperance F, Talajic M. The impact of negative emotions on prognosis following myocardial infarction: is it more than depression? Health Psychol 1995;14:388–98.
41. U.S. Department of Health and Human Services. Mental health: a report of the Surgeon General. Bethesda (MD): National Institute of Mental Health, 1999.
42. Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997;13:641–5.
43. Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995;70: 50–4.
44. Ferencz C, Wiegmann FL Jr., Dunning RE. Medical knowledge of young persons with heart disease. J Sch Health 1980;50:133–6.
45. Anand KJ, Hickey PR. Halothane-morphine compared with high-dose sufentanil for anesthesia and postoperative analgesia in neonatal cardiac surgery. N Engl J Med 1992;326:1–9.
46. Porter FL, Grunau RE, Anand KJ. Long-term effects of pain in infants. J Dev Behav Pediatr 1999;20:253–61.
47. Otterstad JE, Tjore I, Sundby P. Social function of adults with isolated ventricular septal defects: possible negative effects of surgical repair? Scand J Soc Med 1986;14:15–23.
48. Feldman W. How serious are the adverse effects of screening? J Gen Intern Med 1990;5 Suppl:S50–3.
49. Perrin EC. Ethical questions about screening. J Dev Behav Pediatr 1998;19:350–2.
50. Hall S, Bobrow M, Marteau TM. Psychological consequences for parents of false negative results on prenatal screening for Down's syndrome: retrospective interview study. BMJ 2000;320:407–12.

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

John S. Child, MD, FACC, *Co-Chair,* Ruth L. Collins-Nakai, MD, FACC, *Co-Chair*, Joseph S. Alpert, MD, FACC, *Co-Chair*, John E. Deanfield, MD, Louise Harris, MD, ChB, FACC, Peter McLaughlin, MD, FACC, Pamela D. Miner, RN, MN, NP, Gary D. Webb, MD, FACC, Roberta G. Williams, MD, FACC

INTRODUCTION

The expansion of the population of adults with congenital heart disease (CHD) and the increasing survival of patients with complex disease into adulthood have heightened the need for specifically trained individuals who can provide comprehensive outpatient and in-patient care and consultative services to these patients (1–3). In addition, such individuals should have the educational background necessary for successful academic careers in order to advance knowledge and educate other providers. At present, only a few specialists in the U.S. have been specifically trained for this role. Most adult congenital heart disease (ACHD) patients are followed by adult cardiologists who have not had much training in the diagnosis or management of CHD or by pediatric cardiologists who have had little or no experience or training in comprehensive adult care. Adult cardiologists often unofficially consult with pediatric cardiologists to plan management, but uncompensated time and medico-legal risks have made this practice increasingly difficult for pediatric cardiologists. Pediatric cardiologists may effectively co-manage adult patients with an internist or family medicine practitioner, but they cannot provide the full complement of in-patient or invasive services that may be needed. In some cases, adult and pediatric cardiologists follow ACHD patients in a joint clinic. These practices vary considerably depending on patient volume, institutional resources, and physician interest.

The routes by which adult and pediatric cardiologists in this field arrived at their level of expertise are varied. Many, if not most, adult cardiologists have had on-the-job training which provided them with an opportunity to learn, in an environment of collaboration, from pediatric cardiologists and cardiac surgeons. Many pediatric cardiologists have become increasingly involved with adults with CHD as their pediatric patients have aged. Although ACHD patients will continue to rely on these traditionally trained

CL. Growing up with congenital heart disease: the dilemmas of adolescents and young adults. Cardiol Young 1998;8:303–9.
29. Gantt LT. Growing up heartsick: the experiences of young women with congenital heart disease. Health Care Women Int 1992;13: 241–8.
30. Schneider MB, Davis JG, Boxer RA, Fisher M, Friedman SB. Marfan syndrome in adolescents and young adults: psychosocial functioning and knowledge. J Dev Behav Pediatr 1990;11:122–7.
31. Utens EM, Verhulst FC, Erdman RA, et al. Psychosocial functioning of young adults after surgical correction for congenital heart disease in childhood: a follow-up study. J Psychosom Res 1994;38:745–58.
32. Kokkonen J, Paavilainen T. Social adaptation of young adults with congenital heart disease. Int J Cardiol 1992;36:23–9.
33. Van Tongerloo A, De Paepe A. Psychosocial adaptation in adolescents and young adults with Marfan syndrome: an exploratory study. J Med Genet 1998;35:405–9.
34. Alden B, Gilljam T, Gillberg C. Long-term psychological outcome of children after surgery for transposition of the great arteries. Acta Paediatr 1998;87:405–10.
35. Horner T, Liberthson R, Jellinek MS. Psychosocial profile of adults with complex congenital heart disease. Mayo Clin Proc 2000;75:31–6.
36. Casey FA, Sykes DH, Craig BG, Power R, Mulholland HC. Behavioral adjustment of children with surgically palliated complex congenital heart disease. J Pediatr Psychol 1996;21:335–52.
37. Denollet J, Brutsaert DL. Personality, disease severity, and the risk of long-term cardiac events in patients with a decreased ejection fraction after myocardial infarction. Circulation 1998;97:167–73.
38. Kawachi I, Colditz GA, Ascherio A, et al. Prospective study of phobic anxiety and risk of coronary heart disease in men. Circulation 1994; 89:1992–7.
39. Moser DK, Dracup K. Is anxiety early after myocardial infarction associated with subsequent ischemic and arrhythmic events? Psychosom Med 1996;58:395–401.
40. Frasure-Smith N, Lesperance F, Talajic M. The impact of negative emotions on prognosis following myocardial infarction: is it more than depression? Health Psychol 1995;14:388–98.
41. U.S. Department of Health and Human Services. Mental health: a report of the Surgeon General. Bethesda (MD): National Institute of Mental Health, 1999.
42. Kantoch MJ, Collins-Nakai RL, Medwid S, Ungstad E, Taylor DA. Adult patients' knowledge about their congenital heart disease. Can J Cardiol 1997;13:641–5.
43. Cetta F, Warnes CA. Adults with congenital heart disease: patient knowledge of endocarditis prophylaxis. Mayo Clin Proc 1995;70: 50–4.
44. Ferencz C, Wiegmann FL Jr., Dunning RE. Medical knowledge of young persons with heart disease. J Sch Health 1980;50:133–6.
45. Anand KJ, Hickey PR. Halothane-morphine compared with high-dose sufentanil for anesthesia and postoperative analgesia in neonatal cardiac surgery. N Engl J Med 1992;326:1–9.
46. Porter FL, Grunau RE, Anand KJ. Long-term effects of pain in infants. J Dev Behav Pediatr 1999;20:253–61.
47. Otterstad JE, Tjore I, Sundby P. Social function of adults with isolated ventricular septal defects: possible negative effects of surgical repair? Scand J Soc Med 1986;14:15–23.
48. Feldman W. How serious are the adverse effects of screening? J Gen Intern Med 1990;5 Suppl:S50–3.
49. Perrin EC. Ethical questions about screening. J Dev Behav Pediatr 1998;19:350–2.
50. Hall S, Bobrow M, Marteau TM. Psychological consequences for parents of false negative results on prenatal screening for Down's syndrome: retrospective interview study. BMJ 2000;320:407–12.

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

John S. Child, MD, FACC, *Co-Chair,* Ruth L. Collins-Nakai, MD, FACC, *Co-Chair*, Joseph S. Alpert, MD, FACC, *Co-Chair*, John E. Deanfield, MD, Louise Harris, MD, ChB, FACC, Peter McLaughlin, MD, FACC, Pamela D. Miner, RN, MN, NP, Gary D. Webb, MD, FACC, Roberta G. Williams, MD, FACC

INTRODUCTION

The expansion of the population of adults with congenital heart disease (CHD) and the increasing survival of patients with complex disease into adulthood have heightened the need for specifically trained individuals who can provide comprehensive outpatient and in-patient care and consultative services to these patients (1–3). In addition, such individuals should have the educational background necessary for successful academic careers in order to advance knowledge and educate other providers. At present, only a few specialists in the U.S. have been specifically trained for this role. Most adult congenital heart disease (ACHD) patients are followed by adult cardiologists who have not had much training in the diagnosis or management of CHD or by pediatric cardiologists who have had little or no experience or training in comprehensive adult care. Adult cardiologists often unofficially consult with pediatric cardiologists to plan management, but uncompensated time and medico-legal risks have made this practice increasingly difficult for pediatric cardiologists. Pediatric cardiologists may effectively co-manage adult patients with an internist or family medicine practitioner, but they cannot provide the full complement of in-patient or invasive services that may be needed. In some cases, adult and pediatric cardiologists follow ACHD patients in a joint clinic. These practices vary considerably depending on patient volume, institutional resources, and physician interest.

The routes by which adult and pediatric cardiologists in this field arrived at their level of expertise are varied. Many, if not most, adult cardiologists have had on-the-job training which provided them with an opportunity to learn, in an environment of collaboration, from pediatric cardiologists and cardiac surgeons. Many pediatric cardiologists have become increasingly involved with adults with CHD as their pediatric patients have aged. Although ACHD patients will continue to rely on these traditionally trained

JACC Vol. 37, No. 5, 2001
April 2001:1161–98

cardiologists for their care, a specifically trained workforce is called for, as described here.

The aim of this section is to describe the educational requirements for the creation of the specialized cardiology workforce that would be best qualified to fill the roles of caregiver for ACHD patients, team leader of regional programs, and academic leader who will advance the field. The workforce required to successfully care for this population also includes personnel such as experienced mid-level providers (e.g., advanced practice nurses and physician assistants), psychologists, social workers, and obstetricians, but their workforce requirements and educational needs are beyond the scope of this document.

LEVELS OF TRAINING IN ACHD

Because some basic training in CHD is necessary for all adult cardiology trainees in the U.S., a system must be devised that enables adult cardiology training programs to offer educational experience in CHD. At a minimum, this allows the trainee to recognize CHD and attempt to make a preliminary diagnosis, to refer the patient to a regional ACHD center, and to work with that center in the care of these patients.

The terminology used in this document for ascending levels of training (Levels 1, 2, and 3, with Level 3 being the highest) is derived from definitions adopted from the Core Cardiology Training Symposium 2 (COCATS II) (4), which recommends training requirements for adult cardiovascular specialists: Level 1—requires basic training of all adult (medical) cardiology trainees so they may become competent consulting cardiologists; Level 2—requires additional training in a specialized area to enable the cardiologist to perform or interpret, or both, specific procedures or skills at an intermediate skill level; and Level 3—requires additional training in a specialized area to enable the cardiologist to perform, interpret, and train others to perform and interpret specific procedures or acquire skills and knowledge at a high level.

Level 1 training consists of basic exposure to CHD patients and organized educational material on CHD. To enable proper recognition of the problems of adults with CHD, and to be cognizant of when specialized referral is needed, all medical cardiology fellows must achieve Level 1 training in CHD. Level 1 trainees should be instructed by a faculty member with Level 2 or 3 training, or its equivalent. A pediatric cardiologist should also be involved in these training exercises. Level 1 training can be achieved, in part, by core curriculum lectures, assigned reading or audiovisual aids (e.g., videotapes), and case management conferences. Core, or Level 1, training should include didactic material on CHD anatomy, physiology, pathology, genetics, natural history, clinical presentation, and management. Case management conferences should include a review of data on, and medical images of, ACHD patients. During training in electrocardiography, echocardiography, nuclear cardiology, and cardiac catheterization, trainees should be exposed to the evaluation of CHD with these modalities. Postoperative sequelae and residual abnormalities should be stressed, as well as appropriate follow-up protocols and indications for intervention.

Adult cardiology trainees planning to care for ACHD patients (Level 2 training) should have, in addition to the didactic material recommended earlier, at least one year of training in ACHD. This should be an intensive program with exposure to all the components of Level 3 training, but in lesser amounts. Level 3 trainees need at least two years of training.

COMPETENCIES REQUIRED FOR LEVEL 2 AND LEVEL 3 SPECIALISTS

The specific competencies required of Level 3 ACHD leaders and trainers will aid in defining the structure of the training program these individuals will require. They are as follows:

1. Medical and surgical management of CHD.
2. Postoperative management of adults with CHD.
3. Technical and diagnostic expertise in invasive and noninvasive cardiac procedures.
4. Recognition and management of acquired cardiovascular and cardiopulmonary disease.
5. Physiologic changes of pregnancy and awareness of the important effects on and presentation of CHD.
6. Recognition and appropriate initial management of noncardiac disease in adults.
7. Direct and meaningful experience with clinical research methodology, including fundamentals of clinical epidemiology.
8. Embryology, morphology, and pathophysiology of CHD.
9. Principles of health promotion in adults.
10. Psychosocial aspects of adolescence and the transition to adulthood.
11. Recognition of high-risk behaviors in adolescents and adults.
12. Life-style counseling and advocacy for adolescents and adults with CHD.

The cardiologist specializing in ACHD in the U.S. will usually not be fully employed in the care of only ACHD patients and will remain in active practice in either pediatric or adult cardiology. At present, if an adult medical cardiology trainee plans to combine the practice of pediatric and adult CHD, sufficient general pediatric and pediatric cardiology training would be required to attain certification in pediatric cardiology. If a pediatric cardiology trainee wishes to combine the practice of pediatric and adult CHD, sufficient medical cardiology training would be required to qualify him or her for certification in adult cardiovascular medicine.

LEVEL 2 AND LEVEL 3 TRAINING PATHWAYS

The means by which a trainee may arrive at advanced training are currently diverse. No set pattern has been formally recognized for training in this area, but delineation of desirable pathways is appropriate. The time required for the training of future ACHD cardiologists in pediatrics, adolescent medicine, internal medicine, adult and pediatric cardiology, and research methodology should be determined by a special task force of the American Board of Internal Medicine (ABIM) and the American Board of Pediatrics (ABP), facilitated by the American College of Cardiology (ACC). It is strongly recommended that these boards develop some flexibility in the amount of adult combined with pediatric experience required for eligibility for examinations in the future, because of the inordinately long periods currently required for the full complement of training in both fields. There is also the possibility of incorporating the unique Medicine/Pediatrics training program as a pathway to subspecialization in this field.

For Level 2 and 3 trainees, the standards of knowledge and proficiency in echocardiography must include detailed knowledge of all aspects of standard transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) in adults with CHD, in addition to the minimal Level 3 standards for acquired adult heart disease. Echocardiographic training in CHD is usually done best in a pediatric echocardiographic laboratory. Level 3 should be characterized by the ability to independently perform and interpret TTE and TEE studies in a wide range of CHD cases. A minimum of 150 complete TTE and 25 TEE ($\geq$10 intraoperatively) studies of patients with CHD should be performed and interpreted, with participation in the interpretation of at least 300 TTE and 50 TEE studies (20 intraoperatively). The director of the laboratory must make an assessment of each Level 3 trainee's progress and qualifications and thus adapt the number of required studies to the individual. Even for Level 3 cardiology trainees, it is recognized that some aspects of echocardiography (e.g., fetal studies) will not likely fall within their practice skills, and some studies should be referred to a pediatric echocardiographer. Nonetheless, awareness of the role of and implications of fetal echocardiographic data in the management of their ACHD patients is essential.

All cardiology fellows are required to have a defined minimal exposure to cardiac catheterization, including basic knowledge of the various procedures, indications and complications of these procedures, and a specified amount of hands-on training. Level 2 and 3 trainees should also have basic knowledge of the angiographic anatomy of a wide variety of CHD cases, as well as an understanding of the pertinent hemodynamic data. Regular attendance at weekly case management conferences and review of preoperative data, including catheterization and medical imaging data, comprise the first step toward acquiring this familiarity. Direct hands-on catheterization experience in a variety of CHD cases should be required: at least 20 patients for Level 2 and at least 40 patients for Level 3 training in ACHD, over and above core cardiology training (pediatric or adult).

Individuals seeking training in advanced or interventional catheter therapy of ACHD patients will require *at least* one additional year of specialized training at a tertiary care center with large patient volumes and abundant staff expertise. In addition to the need for direct participation in many more diagnostic catheterizations in patients with CHD (minimum of 100 cases) than the minimums described earlier, training should include sufficient exposure to all techniques of CHD interventional therapy, including balloon dilation, vascular stenting, and coil or other device insertion, so that the cardiologist can ultimately be qualified as an independent operator.

Electrophysiology services are vital in managing adults with CHD, particularly in the postoperative group. An electrophysiologist with expertise in ACHD should be involved in the care of any of these patients with recurrent or problematic arrhythmias. Whether such a person is primarily trained in pediatric or adult electrophysiology is not important; however, they should have some training in both pediatric and adult electrophysiology environments.

It is expected that physicians who will be primarily responsible for the management of arrhythmias in ACHD patients will have: 1) Board certification in cardiac electrophysiology (ABIM- or ABP-sponsored Added Qualification Examination); and 2) completed a fellowship training program in adult or pediatric electrophysiology in accordance with North American Society for Pacing and Electrophysiology (NASPE) guidelines (minimum of two years duration). Such individuals will have attained the prerequisite experience in arrhythmia management, pacemaker and automatic implantable cardioverter defibrillator devices, electrophysiology, and intracardiac mapping.

The electrophysiologist caring for adults with CHD must have a sound knowledge of the underlying anatomy and surgical approaches. It would be preferable for such an individual to spend three to six months in a congenital cardiac program (both pediatric and adult) involved in the clinical care of these patients. Such a program should include exposure to the commonly performed surgical procedures. The electrophysiology fellowship program should include a minimum of six months of training specifically in intracardiac mapping and ablation in a recognized center that has substantial expertise in the ablation of complex atrial and ventricular arrhythmias. This would include training in the use of currently available electroanatomic mapping systems. As intra-atrial arrhythmias (as a consequence of underlying pathophysiology and/or previous surgical procedures) are often complex in nature in these patients, and as they are an important contributor to morbidity and possibly to mortality, such training would be essential for the individual wishing to perform catheter ablation in these patients.

JACC Vol. 37, No. 5, 2001
April 2001:1161–98

Level 2 and 3 trainees should be exposed to other specialists working in this area, including cardiac anesthetists, intensivists, other medical subspecialists, mid-level providers, and other professionals, such as psychologists and physical therapists. Level 3 trainees must participate in basic science or clinical research that relates to CHD.

A skilled and versatile cardiovascular surgeon is key in the tertiary care center. A surgeon must have extensive experience in congenital and acquired cardiovascular disorders before acquiring supervised experience in the surgery of ACHD patients. Level 2 and 3 trainees should learn much about the surgical issues that arise in the care of these patients and should be familiar with postoperative problems of common operations, such as repaired tetralogy, atrioventricular septal defect, conduits, and Fontan repair.

Level 2 and 3 trainees should also have a solid understanding of the potential impact of co-morbidities on the patient's management and course; knowledge of the problems of pregnancy in relation to cardiac anomalies and of the effects of drugs on the mother and fetus; and information on contraception, transplantation, exercise, employment, life insurance, and the operation of motor vehicles and airplanes. They should attend regular didactic rounds and case management conferences.

MID-LEVEL PROVIDERS

This term is used to include advanced practice nurses and physician assistants. For mid-level providers and others practicing in an ACHD setting, special training and work experience should usually be built on a strong clinical base in medical or pediatric cardiology. Depending on the needs of the ACHD center, the advanced practice nurse or physician assistant can be specially trained to assist with cardiac catheterizations or to perform echocardiography and other cardio-diagnostic studies. Experience in managing critically ill adults is important, and such personnel may benefit from a background in a coronary care or intensive care unit. Excellent communication skills are imperative. An advanced practice nurse (clinical nurse specialist or nurse practitioner) holds a Master's degree in nursing, with subspecialization in areas such as cardiology, pediatrics, or "acute care." State certification is offered upon graduation from an accredited school. National certification is granted after passing a Board examination. Prescriptive privileges vary from state to state. Physician assistants are certified to practice under their supervising physician's license after a two-year program that prepares them for responsibilities similar to those of advanced practice nurses. Admission to a physician assistant program in the U.S. usually demands a Bachelor's degree, with specific science requirements.

FACILITIES FOR TRAINING IN ADULT CHD

It is likely that Level 2 and 3 training in ACHD will remain the task of tertiary care regional centers over the next decade. A variety of clinical laboratories must provide the trainee with ample exposure to the various techniques employed in caring for adults with CHD: cardiac catheterization, electrophysiology and pacemakers, electrocardiography, exercise and pharmacologic stress testing, Doppler echocardiography, ambulatory electrocardiographic monitoring, nuclear cardiology, magnetic resonance imaging and computed tomography, peripheral vascular testing, pulmonary function, and pathology. In addition, there must be fully equipped cardiac and intensive care units, as well as cardiac and vascular surgery sections. A comprehensive medical library and continuing professional development programs must also be available.

The ACHD team should care for adults with CHD admitted to an in-patient service. To provide a wide range of experience, Level 2 and 3 trainees should participate in the evaluation and management of all adults with CHD admitted to the hospital. In-patient admissions may be for elective or emergency admissions for general medical diseases or conditions related to their malformation (e.g., hemoptysis, endocarditis). Admissions may also be for labor and delivery, diagnostic or interventional catheter procedures, electrophysiologic ablation, or noncardiac surgery. For patients who are admitted directly to the care of a congenital cardiovascular surgeon, a Level 2 or 3 trainee should aid the consulting ACHD cardiologist before and after the operation, as well as provide or arrange any cardiology intraoperative services (e.g., intraoperative transesophageal echocardiography).

RESEARCH AND INTELLECTUAL ENVIRONMENT

A culture of research (from cell to community) needs to be emphasized in ACHD training centers. Clinical research data should be shared through collaborative studies with other centers and peer-reviewed published data should be exchanged with other centers to provide ever-improving care for this group of patients and to enhance the intellectual environment for trainees and faculty. Because each type of complex congenital cardiac disease is relatively rare, necessary information from several tertiary care regional centers should be prospectively pooled to develop clinical studies with sufficient power to answer the research questions, particularly those examining interventions and outcomes.

The creation of specific ACHD research fellowships is recommended. This would permit individuals to dedicate 75% to 100% of their time to research, over a guaranteed two- to three-year period, any aspect of ACHD (biomedical, clinical, health services, or population research). Such fellowships would help build a cadre of enhanced research personnel in ACHD. It is further recommended that a specific network of centers of excellence in care for adults with CHD be created and funded through the National Heart, Lung, and Blood Institute.

CONCLUSIONS

At present, the physician workforce caring for ACHD patients in the U.S. consists of a few (<20) adult cardiologists with advanced training, as described, and an ongoing career focus in ACHD, as well as a much larger number of adult and pediatric cardiologists with little or no specific training in the care of ACHD patients, but with on-the-job experience. Development of a small but highly trained cohort of ACHD specialists who could lead an integrated network of specialized centers would improve clinical care, advance knowledge, and help provide ongoing professional education for the larger population of adult and pediatric cardiologists who care for the majority of these patients.

Creating this population of ACHD specialists requires the clear articulation of training pathways and certification. Because of the long time required for training in CHD and adult diseases and research, some consolidation of training will be needed, in addition to the development of specific training funds and the establishment of debt relief to attract and maintain an adequate workforce.

RECOMMENDATIONS

- A joint task force of the ABIM and ABP, facilitated by the ACC, should be formed to determine the specific pathways and years of training required for Level 2 and 3 ACHD subspecialist cardiologists.
- Level 2 and 3 training programs should be coordinated to ensure the greatest learning opportunities for the ACHD cardiologists-in-training and to provide continuing education for trainees, graduates, and ACHD practitioners.
- ACHD research fellowships should be created so that individuals can spend 75% to 100% of their time in protected research over a two- to three-year period.
- Training programs for other key staff (e.g., nurses, physician assistants, psychologists, social workers, other nonphysician personnel) on ACHD teams should be established.

TASK FORCE 3 REFERENCE LIST

1. Congenital heart disease after childhood: an expanding patient population. Presented at the 22nd Bethesda Conference, Bethesda, Maryland, October 18–19, 1990. J Am Coll Cardiol 1991;18:311–42.
2. Perloff JK. Congenital heart disease in adults: a new cardiovascular subspecialty. Circulation 1991;84:1881–90.
3. Perloff JK, Miner PD. Specialized facilities for the comprehensive care of adults with congenital heart disease. In: Perloff JK, Child JS, editors. Congenital Heart Disease in Adults. Philadelphia: W. B. Saunders, 1998:9–12.
4. Beller GA, Bonow RO, Fuster V, et al. Guidelines for training in adult cardiovascular medicine: Core Cardiology Training Symposium (COCATS II). J Am Coll Cardiol. In press.

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

Michael J. Landzberg, MD, FACC, *Co-Chair*, Daniel J. Murphy, Jr, MD, FACC, *Co-Chair*, William R. Davidson, Jr, MD, FACC, John A. Jarcho, MD, FACC, Harlan M. Krumholz, MD, FACC, John E. Mayer, Jr, MD, FACC, Roger B. B. Mee, MD, ChB, David J. Sahn, MD, FACC, George F. Van Hare, MD, FACC, Gary D. Webb, MD, FACC, Roberta G. Williams, MD, FACC

ORGANIZATION OF DELIVERY SYSTEMS FOR ACHD

The delivery of appropriate care to adults with congenital heart disease (ACHD) is a largely unmet challenge in the U.S. and elsewhere. To meet this challenge, a structure and process for the organization and delivery of care is proposed. We will use the "severe heart failure care model" familiar to most cardiologists as an example of how the needs of ACHD patients can best be met. Similar to the challenge of the severe heart failure patients, ACHD patients have a low-to-moderate prevalence, need caregivers with both special knowledge of the conditions encompassed and the ability to provide tailored and out-of-the-ordinary treatments, and may require high-intensity medical care. By contrast to the heart failure population, ACHD patients reach age 18 at a rate of about 9,000 annually in the U.S. and may require much longer surveillance and care than most heart failure patients.

In this section we will: 1) describe the "severe heart failure model" that we propose should be emulated for ACHD patients, 2) describe the structure of such a program based on the concept of regional ACHD centers across the U.S., 3) outline the resources (services and personnel) required in such centers, 4) propose responsibilities for different types of physicians in the care of these patients, 5) describe the initial patient visit and its goals, 6) propose strategies for long-term follow-up, 7) and make some comments regarding hospitalization of these patients.

SEVERE HEART FAILURE AS A MODEL OF REGIONALIZATION AND CENTRALIZATION

The established "local caregiver or center supported by a regional specialized center" model for the organization and delivery of care for adult patients with severe heart failure serves as a paradigm for our proposal for a system of care for ACHD. When compared with the average cardiology patient, those with severe heart failure tend to carry high

CONCLUSIONS

At present, the physician workforce caring for ACHD patients in the U.S. consists of a few (<20) adult cardiologists with advanced training, as described, and an ongoing career focus in ACHD, as well as a much larger number of adult and pediatric cardiologists with little or no specific training in the care of ACHD patients, but with on-the-job experience. Development of a small but highly trained cohort of ACHD specialists who could lead an integrated network of specialized centers would improve clinical care, advance knowledge, and help provide ongoing professional education for the larger population of adult and pediatric cardiologists who care for the majority of these patients.

Creating this population of ACHD specialists requires the clear articulation of training pathways and certification. Because of the long time required for training in CHD and adult diseases and research, some consolidation of training will be needed, in addition to the development of specific training funds and the establishment of debt relief to attract and maintain an adequate workforce.

RECOMMENDATIONS

- A joint task force of the ABIM and ABP, facilitated by the ACC, should be formed to determine the specific pathways and years of training required for Level 2 and 3 ACHD subspecialist cardiologists.
- Level 2 and 3 training programs should be coordinated to ensure the greatest learning opportunities for the ACHD cardiologists-in-training and to provide continuing education for trainees, graduates, and ACHD practitioners.
- ACHD research fellowships should be created so that individuals can spend 75% to 100% of their time in protected research over a two- to three-year period.
- Training programs for other key staff (e.g., nurses, physician assistants, psychologists, social workers, other nonphysician personnel) on ACHD teams should be established.

TASK FORCE 3 REFERENCE LIST

1. Congenital heart disease after childhood: an expanding patient population. Presented at the 22nd Bethesda Conference, Bethesda, Maryland, October 18–19, 1990. J Am Coll Cardiol 1991;18:311–42.
2. Perloff JK. Congenital heart disease in adults: a new cardiovascular subspecialty. Circulation 1991;84:1881–90.
3. Perloff JK, Miner PD. Specialized facilities for the comprehensive care of adults with congenital heart disease. In: Perloff JK, Child JS, editors. Congenital Heart Disease in Adults. Philadelphia: W. B. Saunders, 1998:9–12.
4. Beller GA, Bonow RO, Fuster V, et al. Guidelines for training in adult cardiovascular medicine: Core Cardiology Training Symposium (COCATS II). J Am Coll Cardiol. In press.

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

Michael J. Landzberg, MD, FACC, *Co-Chair*, Daniel J. Murphy, Jr, MD, FACC, *Co-Chair*, William R. Davidson, Jr, MD, FACC, John A. Jarcho, MD, FACC, Harlan M. Krumholz, MD, FACC, John E. Mayer, Jr, MD, FACC, Roger B. B. Mee, MD, ChB, David J. Sahn, MD, FACC, George F. Van Hare, MD, FACC, Gary D. Webb, MD, FACC, Roberta G. Williams, MD, FACC

ORGANIZATION OF DELIVERY SYSTEMS FOR ACHD

The delivery of appropriate care to adults with congenital heart disease (ACHD) is a largely unmet challenge in the U.S. and elsewhere. To meet this challenge, a structure and process for the organization and delivery of care is proposed. We will use the "severe heart failure care model" familiar to most cardiologists as an example of how the needs of ACHD patients can best be met. Similar to the challenge of the severe heart failure patients, ACHD patients have a low-to-moderate prevalence, need caregivers with both special knowledge of the conditions encompassed and the ability to provide tailored and out-of-the-ordinary treatments, and may require high-intensity medical care. By contrast to the heart failure population, ACHD patients reach age 18 at a rate of about 9,000 annually in the U.S. and may require much longer surveillance and care than most heart failure patients.

In this section we will: 1) describe the "severe heart failure model" that we propose should be emulated for ACHD patients, 2) describe the structure of such a program based on the concept of regional ACHD centers across the U.S., 3) outline the resources (services and personnel) required in such centers, 4) propose responsibilities for different types of physicians in the care of these patients, 5) describe the initial patient visit and its goals, 6) propose strategies for long-term follow-up, 7) and make some comments regarding hospitalization of these patients.

SEVERE HEART FAILURE AS A MODEL OF REGIONALIZATION AND CENTRALIZATION

The established "local caregiver or center supported by a regional specialized center" model for the organization and delivery of care for adult patients with severe heart failure serves as a paradigm for our proposal for a system of care for ACHD. When compared with the average cardiology patient, those with severe heart failure tend to carry high

levels of medical complexity and incidence of recurrent illness, and they have less-optimal outcomes.

Given the supposition by internists and cardiologists that a great deal of heart failure management falls within their own expertise, patient care, including that for the most severely ill, previously tended to be spread throughout all levels of adult cardiovascular care. This model tended to limit the capacity to expand services, apply new knowledge, share experiences, and compare outcomes. The organization of best practice guidelines was difficult, and translation of such recommendations to everyday care was limited. Improvement in average care was gradual.

Because of a growing accountability to third-party payers and limited organ donor procurement, a new model for organizing and delivering care to the most severely ill arose, centered around a specialized regional program and working in conjunction with local providers of care. This system has evolved over a 20-year period, fulfilling most expectations for the provision of high-quality care. The severe heart failure model has allowed for: 1) improved teaching, collection, and dissemination of knowledge regarding heart failure and its ramifications; 2) new treatments, many of which could not be tested without sufficient numbers of patients and resources; 3) decreased outpatient visits, fewer hospitalizations, and improved patient quality of life; 4) improved medical and surgical outcomes; 5) containment of costs; 6) a more uniform pattern of medical care (allowing improved cooperation and cross-referral of patients and better definition of the appropriateness of medical and surgical care at a local, compared with a regional, center); and 7) a greater interaction between third-party payers, insurers, and medical caregivers.

This model has required the growth and development of both a national registry and regional databases to collect, organize, interpret, and distribute standardized and requested information and to review this in a timely fashion. Individual institutions maintain financial commitments to the maintenance of the databases and to the employment of medically savvy data collection and entry personnel. All institutions have access to their individual data and have the opportunity to initiate issue-driven research. Evidence-based recommendations can be generated with actual data and analysis requested by and determined in large part by the medical caregivers themselves.

The local and regional model of medical care functions well for this relatively small group of patients in need of expert and evidence-based care. A similar system will allow caregivers for ACHD to achieve the same rewards already obtained for adults with severe heart failure.

EVALUATION OF QUALITY

Health care quality has been classified into three components: structure (training and skills of personnel, adequacy of diagnostic and therapeutic equipment resources, and organizational systems that mobilize these resources most efficiently for optimal patient care), process (the use of appropriate diagnostic and therapeutic modalities for individual patients), and outcomes (the consequences of treatment).

PROPOSED STRUCTURE OF THE HEALTH CARE DELIVERY SYSTEM FOR ADULTS WITH CONGENITAL HEART DISEASE

An algorithm for the initial evaluation and ongoing care of ACHD is proposed. These recommendations include the subdivision and coordination of care of ACHD both locally and at regional ACHD centers. This model requires a system of data storage, rapid communication, critical self-analysis, establishment and implementation of practice guidelines, and insights to provide for the coordination of optimal current and future care of ACHD.

LOCAL (INDIVIDUAL PHYSICIAN AND CARDIOLOGIST)

Local medical resources for ACHD may be a family doctor, an internist, or a general cardiologist on the one hand, and an ACHD cardiologist with a commitment to, training in, and/or experience with the care of ACHD on the other. The first three groups of physicians will usually have a major or exclusive role in the types of patients listed in Table 6 of Task Force #1. These local clinicians might also participate in the care of adults with moderate and complex CHD (Tables 4 and 5 of Task Force #1) in collaboration with the staff of a regional ACHD center.

The ACHD cardiologists (who also practice as pediatric or adult medical cardiologists) can care for any ACHD patient. At present, the majority of ACHD cardiologists will have had informal training and on-the-job experience in the care of ACHD (see Task Force #3). More recently, a few training centers have produced ACHD cardiologists with comprehensive training and often a commitment to contribute academically to the ACHD discipline.

THE REGIONAL ACHD CENTER

A regional ACHD center is usually directed by an ACHD cardiologist who is supported by a collaborative, multidisciplinary team involving other cardiologists, mid-level practitioners, congenital heart surgeons, and others. The specific components of such a program are outlined in Table 1. Regional ACHD centers will frequently serve as the entry point for ACHD. They may receive patients from sources such as general pediatric and adult medical cardiologists, other specialists (e.g., obstetricians), primary care providers, patient self-referrals, and medical insurers. Every ACHD patient should be evaluated at least once by an ACHD cardiologist for the purpose of initial evaluation and recommendations for long-term care. Ideally, this applies even to the patients in Table 6 of Task Force #1, so-called simple CHD. This is particularly true for patients who have not been under the care of pediatric cardiologists. The goal of

Table 1. Personnel and Services Recommended or Required for Regional ACHD Centers

Type of Service or Personnel	Local Care	Regional ACHD Center
Pediatric ACHD cardiologist	Optional	One or several 24/7*
Adult medical ACHD cardiologist	Optional	One or several 24/7*
Mid-level practitioner	Optional	Two/several
Congenital heart surgeon	No	Two/several 24/7*
Cardiac anesthesia	No	Several 24/7*
Echocardiography** Includes TEE, intraoperative TEE (required for surgery)	Refer to regional ACHD center	Two/several 24/7*
Diagnostic catheterization**	Refer to regional ACHD center	Yes 24/7*
Noncoronary interventional catheterization**	Refer to regional ACHD center	Yes 24/7*
Electrophysiology**	Consult regional ACHD center unless unrelated to CHD	Yes 24/7*
Exercise testing	Standard	Echo, radionuclide, cardiopulmonary, metabolic
Transplant	Optional	Heart, lung, heart-lung desirable
Cardiac imaging/radiology services	Optional	CT scan, cardiac MRI with fast-pulse sequencing*, nuclear medicine
Cardiac pathology	Optional	Yes
Information technology	Optional • Interface with regional ACHD center • Data collection • Participation in patient-care and best-practice guidelines review	• Data collection • Database support • Interface with local practitioners, including internet-based applications • Quality assessment review and protocols • Optional development of best practice guidelines
Other		• Adolescent transitional unit • High risk obstetrics • Genetics • Rehabilitation services • Social services • Vocational services • Financial counselors

* "24/7" denotes availability 24 hours/day, 7 days/week. **These modalities must be supervised/performed and interpreted by physicians with specific skills and knowledge in CHD, as outlined.

the visit is to ensure that other diagnoses or subtle but important findings have not been missed. Too often, patients with "simple" CHD are seen who have been misdiagnosed, mismanaged, or misinformed. Caregiver and insurance referral patterns will often require reconfiguration for referral to caregivers with specific expertise in ACHD care.

Regional ACHD centers may be established within an adult hospital, a children's hospital, a unit shared by both children and adult hospital facilities, or a freestanding unit. Such centers must afford prompt access for patients and referring physicians in order to provide:

Comprehensive diagnosis—All modes of cardiac diagnosis should be available. Each component of the diagnostic evaluation should be performed by individuals with appropriate training and experience in CHD.

Management planning—Best decisions have traditionally occurred within the venue of a case-management conference, at which personnel from cardiology, cardiac surgery, anesthesia, intensive care, and nursing review relevant data. Case-management conferences with discussion and consensus are very important in determining care strategy (including both the nature and timing of intervention) as well as educating and building the cohesion of team members.

Patient counseling—Within a regional ACHD center adults with CHD should participate in an informed discussion of their current medical/cardiac situation and their proposed management plan.

Specific personnel and services within regional ACHD centers are also necessary, including:

Cardiac anesthesia—The presence of a cardiac anesthesia team that offers consultative services, interacts with other members of the ACHD caregiving team, and anesthetizes patients with CHD is required.

Operating rooms—Operating facilities with prompt or immediate access to all perioperative (e.g., echocardiography, catheterization) and intraoperative (e.g., transesophageal echocardiography) diagnostic procedures are essential. Dedicated fully trained congenital cardiac perfusionists (with expertise in VAD and ECMO setup, delivery, and maintenance) are mandatory.

Cardiac surgery—In addition to adult cardiovascular surgeons, regional ACHD centers require the availability of full-time, expert congenital heart surgeons. At least two congenital heart surgeons (often based primarily at a children's hospital) are required to provide 24-h coverage for both the pediatric and adult facilities. Their surgical teams should be expected to maintain their expertise through performing a critical annual volume of pediatric and ACHD surgeries.

Intensive Care—ICU staff trained and expert in provision of care to ACHD are required in regional ACHD centers. The ICU should be sited with rapid access to the ORs and be capable of performing open-chest resuscitation and of implementing and monitoring ECMO and VAD. The ICU staff and residents/fellows can be culled from medical cardiology, cardiac anesthesia, cardiac surgery, and critical care specialties, and they should be supported by fellowship programs. Expert medical and surgical care should be on-site 24 h/day, 7 days/week. The skill of the staff in diagnosing and managing acquired cardiovascular and other diseases is very important here as well as throughout all units and services caring for ACHD patients. Timely access to all diagnostic services and interventions should be available 24 h/day. The ICU nursing staff should have specific expertise in the care and management of ACHD.

In-patient service—ACHD patients require a hospital environment with specifically qualified nursing staff and support personnel. This may be provided within the context of other medical or cardiology unit or on a unit dedicated to ACHD patient care. The unit should contain a high-intensity central nursing area with hemodynamic/electrocardiographic telemetry monitoring. Expert medical and surgical physician care should be either on-site or available in a near-immediate fashion 24 h/day, 7 days/week. Optimally, the in-hospital beds, ICU, cath lab, and ORs should be geographically clustered, in close proximity to noninvasive laboratories, outpatient areas, and cardiology/cardiac surgery administrative services. The center should support social workers and financial counselors, and it should make appropriate use of chaplaincy support.

Transplantation—Regional ACHD centers should be affiliated with a transplant program.

Catheterization laboratory—The provision of expert diagnostic and therapeutic cardiac catheterization skills for ACHD requires personnel specifically trained in ACHD and needed therapies as well as all aspects of adult acquired medical disease. Table 2 describes the types of patients who should have cardiac catheterizations performed in regional ACHD centers. The catheterization laboratory and its equipment, as well as the recovery and post-catheterization ward facilities, must be provided. Finally, to maintain excellence, the laboratories and personnel at regional ACHD centers should have continuous experience at sufficient levels of adult or pediatric CHD complexity and volume.

Noninvasive imaging service—24 h/day, 7 day/week coverage is required, with volume and complexity sufficient to maintain excellence in obtaining and interpreting echocardiographic, computerized tomographic, and magnetic resonance images of ACHD patients.

Electrophysiology service—A fully equipped and properly staffed service with a full range of ablative and pacing therapies, in addition to the consultative and diagnostic services appropriate to the special needs of ACHD patients, must be available.

Table 2. Types of Patients Needing Cardiac Catheterization in Regional ACHD Centers

The following cardiac catheterization procedures on ACHD should be performed at a regional ACHD center and by staff with sufficient training, expertise and support services (including congenital heart surgical backup):
All diagnostic catheter procedures with the exception of coronary angiography
Aortic coarctation/RV outflow/peripheral pulmonary stenosis dilation &/or stent placement
Congenital valve dilation
PDA closure
Septal defect closure
Vasodilator or vascular shunt/access occlusion trials
Venous pathway dilation or stent placement

High-risk obstetrics—24 h/day, 7 day/week coverage by staff expert in the counsel and care of women with CHD is a special requirement.

Cardiac pathology—Expertise in congenital cardiac pathology and post-mortem examination must be available within the regional ACHD centers.

GEOGRAPHIC DISTRIBUTION OF REGIONAL ACHD CENTERS

The proposed regionalization described in this report should provide appropriate and continuous access, when needed, to all types of care for all ACHD in the U.S. Because geographic regions of the U.S. vary in population density and available medical resources, some flexibility in applying the principles of regionalization is appropriate. As a rule, there should be approximately one regional ACHD center per population of 5 to 10 million people and approximately 30 to 50 regional ACHD centers nationwide. In some areas of the country, regional ACHD centers may be farther apart and may have somewhat smaller ACHD populations. In the largest urban centers with several pediatric cardiology and congenital heart surgical programs there are likely to be two or more regional ACHD centers. In all regions, reciprocal communication between regional ACHD centers, local caregivers, and patients is required. In recognition of the fact that particular areas of expertise may not be equally present in each regional ACHD center, specific geographic referral patterns may overlap different regions.

PROPOSED PROCESS FOR DELIVERY OF HEALTH CARE TO ACHD PATIENTS

Newly arrived ACHD patients. As described in the report of Task Force #2, an orderly transition of care from the pediatric to the adult facility is most strongly recommended. One of the many reasons for this is to reduce the number of patients lost to follow-up during adolescence and young adult life. The pediatric cardiologist should provide a copy of all relevant clinical records, including operative reports, catheterization reports, and the like, to the patients and the regional ACHD centers at the time of transfer to ACHD care.

The initial patient evaluation. Patients may first present for CHD care in their adult years because they have new symptoms, functional deterioration, or a growing sense of the need to resume regular care.

An ACHD specialist should evaluate all adults with moderate and complex CHD (Tables 4 and 5 of Task Force #1) at least once and should also evaluate most patients with simple CHD (Table 6 of Task Force #1). The evaluation should include a thorough history, a review of documents outlining specific diagnoses and details of treatments applied, and any other clinical problems. In addition, a tailored clinical and laboratory evaluation should be performed to assess current patient status. This initial ACHD evaluation should also involve an extensive component of patient education regarding both the nature of the congenital abnormality and the anticipated unrepaired or postoperative course, along with instructions on when and how to access care in the future, especially in urgent situations. This consultation should result in a report to patients and their primary care and supporting physicians. This report will document the baseline evaluation and provide a contact for questions and other issues that may arise in the future. The initial ACHD evaluation allows stratification of these patients according to risk and management difficulty.

An ACHD cardiologist will review the history regarding acquired cardiovascular and other medical conditions. This should be part of each work-up and will increase in importance as a patient ages. For example, the development of coronary artery disease or high blood pressure is important not only in itself but also in its potentially adverse effect on the course of CHD in adults.

Long-term follow-up. Most ACHD patients will require intermittent regular evaluations at a regional ACHD center. Such patients will benefit by maintaining contact with a primary care physician and, in some cases, a local adult medical cardiologist. All reports generated at regional ACHD centers should be transmitted to patients and their local physicians and should include specific goals and responsibilities for local as well as regional ACHD follow-up. In some cases, when a patient lives close to a regional ACHD center, the ACHD cardiologist can function as a primary cardiologist, leaving other health care to the primary care physician.

It is not implied here that the regional ACHD center take over the care of all ACHD patients. The role of the regional center should be to take an appropriate role in the management of each patient (ranging from no role, through joint care, to exclusive and close care). In addition, it should be used as a source of expert advice and information.

For simplicity, three groups of patients are described according to the following scheme:

Lesions that can usually be cared for in the Community (Table 6 of Task Force #1) after initial expert evaluation, usually in a regional ACHD center. These patients with simple CHD are felt to be at low risk for new clinical problems. This group includes some patients with minor congenital abnormalities who have not undergone surgical or other intervention (e.g., mild pulmonary valve stenosis, small isolated ventricular septal defect) and patients with simple congenital defects who have undergone successful repair (e.g., repaired ductus arteriosus, ventricular septal defect or secundum atrial septal defect with no residual shunt or other sequelae). Patients in this category will usually be followed by either a primary care physician or a community cardiologist. If necessary, a patient could be referred to a regional ACHD center.

Adults with CHD with residual hemodynamic or structural abnormalities who are clinically stable (Tables 4 and 5 of Task Force #1). Most adults with moderate and complex CHD fall into this category. Each specific defect or combination of defects carries its own list of potential complications. Such patients require ongoing surveillance to detect any changes in status and/or increased risk profile. In addition, as clinical practice and research advance, new principles of patient management will be applied by the ACHD cardiologist at the regional ACHD center. Such patients benefit, as well, from care given by a primary caregiver who provides local ongoing care and who communicates and cooperates with the ACHD cardiologist. For some patients, clinical evaluations may alternate between the local provider and the regional ACHD center.

Adults with CHD may develop active cardiovascular problems or become clinically unstable. These problems should be addressed, whenever possible, at a regional ACHD center. The ACHD cardiologist should maintain primary clinical responsibility for these patients until their clinical status stabilizes. Examples include significant arrhythmias, ventricular dysfunction, significant valve regurgitation, and infective endocarditis. Interventions in such patients generally should be performed at regional ACHD centers.

Any adult with CHD who develops a new clinical problem that might be related to a cardiovascular abnormality should be referred for re-evaluation to, or be under the care of, a regional ACHD center. In addition, if intervention is required, most patients should be evaluated at their regional ACHD center before intervention. When appropriate, some procedures can be performed locally (for example, noncardiac surgery in an asymptomatic low-risk adult with CHD). Such an evaluation might also lead to a recommendation that the intervention be performed at a regional facility integrated with the regional ACHD center.

FREQUENCY OF PATIENT FOLLOW-UP

For adults with CHD in the lowest risk group (Table 6 of Task Force #1), routine cardiac follow-up is recommended every three to five years as a rule.

The larger group of adults with moderate and complex CHD (Tables 4 and 5 of Task Force #1) requires more frequent follow-up, generally every 12 to 24 months. Such evaluation should include a detailed history and clinical

examination. Diagnostic studies should be standardized, with performance of more extensive evaluations (e.g., cardiopulmonary/metabolic stress testing, cardiac MRI, cardiac catheterization) based on the individual patient's clinical course and findings. Part of such evaluations should include the detection of any new or progressive cardiac problems, patient education, and education of the primary care physician.

Finally, a smaller group of adults with CHD with complex anatomy and physiology require serial follow-up and examination at a regional ACHD center every 6 to 12 months, if not more frequently. This patient group includes adult patients with conditions such as single ventricle physiology, a morphologic right ventricle functioning in the systemic circuit, recalcitrant heart failure, recurring arrhythmias, and pulmonary vascular obstructive disease.

URGENT/EMERGENCY CARE

Most adults with CHD should wear medical alert devices and/or carry on their persons information that focuses on issues such as major diagnoses, the use of prosthetic valves or devices, anticoagulation, or other key points.

Emergency medical personnel at regional ACHD centers must be able to provide acute care for adults with CHD. The following situations and conditions go beyond the routine competence of many ER physicians and surgeons: intracardiac or intravascular shunts, pulmonary vascular disease, right ventricular dysfunction, and high-risk pregnancy.

Hospitalization for medical or cardiac acute care. Adults with moderate or severe CHD will usually require transfer to a regional ACHD center for urgent or acute care. This group includes patients with:

- Important intracardiac shunting;
- Greater than "mild" pulmonary vascular disease;
- Greater than "moderate" left ventricular or "mild" right ventricular dysfunction or failure;
- A systemic right ventricle;
- Single ventricle physiology;
- Greater than "mild" obstructive intracardiac valvular or vascular disease, including peripheral pulmonary artery stenosis or aortic coarctation, and excluding isolated aortic valve and many isolated mitral valve patients;
- Important congenital coronary arterial abnormalities;
- Pregnancy in the setting of important CHD;
- New onset of symptomatic tachyarrhythmias requiring institution of antiarrhythmic medication or ablation therapy, or bradyarrhythmias that include AV block or symptomatic sinus node dysfunction, in any of the patients listed above, repaired or unrepaired.

Patients with milder forms of CHD can usually receive their in-patient care in their community, sometimes in consultation with the specialized ACHD regional center. Representative examples include:

- Minimal residual intracardiac/vascular shunting with good ventricular function
- ASD, VSD, PDA corrected with good hemodynamic result
- New onset of symptomatic tachyarrhythmias requiring institution of antiarrhythmic medication or ablation therapy, or bradyarrhythmias that include AV block or symptomatic sinus node dysfunction, in patients with well-repaired ASD, VSD, or AV septal defect.

Non-emergent hospitalization should be based on the same general principles outlined above. Patients with moderate and complex lesions will often require longer and more costly admissions than other types of patients.

INTERVENTIONS

The increasing complexity and procedural requirements for adults with CHD is reflected in their greater than 60% prevalence of prior cardiac operations and their nearly 50% need for re-operation or interventional therapy at some point during adulthood. A review of hospitalizations over the past five years in one center with particular expertise in catheterization of adults with CHD revealed that 26% are non-procedural, 57% involve catheterization and 17% involve surgery. The unique and increasingly complex needs of adults with CHD mandates centralization of procedural care.

TREATMENT OUTCOMES

The evaluation of structure and process requires that the best approach be determined. Ideally, this determination should be based on strong evidence. Expert consensus is necessary when evidence is lacking, but it should not be considered a fair substitute for rigorously performed clinical studies. The field of ACHD faces substantial challenges in generating the evidence needed to define what the "best practices" are. Patient groups are heterogeneous both between and within disease categories. The numbers of patients within particular categories of CHD tend to be small. The need for long-term follow-up in assessing clinical outcomes will delay the evaluation of the effects of new technologies and treatments.

The measurement of outcomes is an appropriate indicator of quality because it is the composite result of what is achieved with both structure and process. Outcomes should be systematically tracked, evaluated, and improved; and outcome data can be used to identify opportunities to improve practice.

Caregivers for adults with CHD, in coordination with third-party payers and regulators of access to health care, have a unique opportunity to construct and effectively utilize data sources, in concert with other non-caregiver-established databases (e.g., Medicare). In such a fashion, questions asked by patient advocacy groups, caregivers, and payer/insurers concerning optimal care strategies and esti-

mates of resource needs and utilization can be effectively addressed.

RECOMMENDATIONS

- Care of adults with CHD should be coordinated by regional ACHD centers that represent a resource for the medical community.
- An individual primary caregiver or cardiologist without specific training and expertise in adult CHD should manage adults with moderate and complex CHD only in collaboration with a physician with advanced training and experience in caring for adults with CHD.
- Every academic adult cardiology/cardiac surgery center should have access to a regional ACHD center for consultation and referral.
- Every cardiologist should have a referral relationship with a regional ACHD center.
- Approximately one regional ACHD center should be created to serve a population of 5 to 10 million people, with 30 to 50 such centers in the U.S.
- Within a single urban center, institutions should establish collaborative relationships.
- Each pediatric cardiology program should identify the ACHD center to which the transfer of patients will be made.
- An ACHD specialist should evaluate all adults with moderate and complex CHD at least once. The initial ACHD evaluation allows stratification of these patients according to risk and management difficulty.
- Adults with moderate and complex CHD will require regular evaluations at a regional ACHD center and will benefit from maintaining contact with a primary care physician.
- For adults with CHD in the lowest risk group (simple CHD), cardiac follow-up is recommended at least every three to five years. The larger group of adults with moderate and complex CHD will require more frequent follow-up, generally every 12 to 24 months. A smaller group of adults with very complex or unstable CHD will require follow-up at a regional ACHD center at a minimum of every 6 to 12 months.
- Every adult with CHD should have a primary care physician. To ensure communication, current clinical records should be on file both at a regional ACHD center and with the primary care provider (patients should also have copies of relevant records).
- All emergency care facilities should have an affiliation with a regional ACHD center.
- Patients with moderate or complex CHD require admission or transfer to a regional ACHD center for urgent or acute care.
- Most cardiac catheterization and electrophysiology procedures for adults with moderate and complex CHD should be performed in a regional ACHD center with appropriate experience in CHD, and in a laboratory with appropriate personnel and equipment. After consultation with staff in regional ACHD centers, it may be appropriate for local centers to perform such procedures.
- Surgical procedures in adults with CHD as outlined in Tables 4 and 5 of Task Force #1 should generally be performed in a regional ACHD center with specific excellence in the surgical care of CHD.
- Each regional ACHD center should participate in a medical and surgical database aimed at defining and improving outcomes in adults with CHD.
- Each regional ACHD center should encourage all ACHD patient data to be included in a national CHD database. Programs should work collaboratively on multicenter projects and develop investigator-initiated research proposals dealing with ACHD.
- The American College of Cardiology should recommend to the NHLBI and/or Agency for Health Care Research and Quality the formation of adult congenital centers for documenting and improving outcomes, education, and research.
- Each regional ACHD center should establish or affiliate with a patient advocacy group.

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

David J. Skorton, MD, FACC, *Co-Chair*, Arthur Garson, Jr., MD, MPH, MACC, *Co-Chair*, Hugh D. Allen, MD, FACC, James M. Fox, MS, MD, Susie C. Truesdell, PA, MBA, Gary D. Webb, MD, FACC, Roberta G. Williams, MD, FACC

INTRODUCTION

Access to optimal, specialized, appropriate health care, health and life insurance, and full employment remains a problem for many adolescent and adult patients with congenital heart disease (CHD) (1).

Health insurance may be difficult to obtain in adulthood because of "pre-existing conditions"—despite recent federal legislation—and because of uncertainties and misconceptions about the cost of care for adults with CHD. The actual costs of medical care appear to be relatively low in these patients compared with survivors of other chronic diseases

mates of resource needs and utilization can be effectively addressed.

RECOMMENDATIONS

- Care of adults with CHD should be coordinated by regional ACHD centers that represent a resource for the medical community.
- An individual primary caregiver or cardiologist without specific training and expertise in adult CHD should manage adults with moderate and complex CHD only in collaboration with a physician with advanced training and experience in caring for adults with CHD.
- Every academic adult cardiology/cardiac surgery center should have access to a regional ACHD center for consultation and referral.
- Every cardiologist should have a referral relationship with a regional ACHD center.
- Approximately one regional ACHD center should be created to serve a population of 5 to 10 million people, with 30 to 50 such centers in the U.S.
- Within a single urban center, institutions should establish collaborative relationships.
- Each pediatric cardiology program should identify the ACHD center to which the transfer of patients will be made.
- An ACHD specialist should evaluate all adults with moderate and complex CHD at least once. The initial ACHD evaluation allows stratification of these patients according to risk and management difficulty.
- Adults with moderate and complex CHD will require regular evaluations at a regional ACHD center and will benefit from maintaining contact with a primary care physician.
- For adults with CHD in the lowest risk group (simple CHD), cardiac follow-up is recommended at least every three to five years. The larger group of adults with moderate and complex CHD will require more frequent follow-up, generally every 12 to 24 months. A smaller group of adults with very complex or unstable CHD will require follow-up at a regional ACHD center at a minimum of every 6 to 12 months.
- Every adult with CHD should have a primary care physician. To ensure communication, current clinical records should be on file both at a regional ACHD center and with the primary care provider (patients should also have copies of relevant records).
- All emergency care facilities should have an affiliation with a regional ACHD center.
- Patients with moderate or complex CHD require admission or transfer to a regional ACHD center for urgent or acute care.
- Most cardiac catheterization and electrophysiology procedures for adults with moderate and complex CHD should be performed in a regional ACHD center with appropriate experience in CHD, and in a laboratory with appropriate personnel and equipment. After consultation with staff in regional ACHD centers, it may be appropriate for local centers to perform such procedures.
- Surgical procedures in adults with CHD as outlined in Tables 4 and 5 of Task Force #1 should generally be performed in a regional ACHD center with specific excellence in the surgical care of CHD.
- Each regional ACHD center should participate in a medical and surgical database aimed at defining and improving outcomes in adults with CHD.
- Each regional ACHD center should encourage all ACHD patient data to be included in a national CHD database. Programs should work collaboratively on multicenter projects and develop investigator-initiated research proposals dealing with ACHD.
- The American College of Cardiology should recommend to the NHLBI and/or Agency for Health Care Research and Quality the formation of adult congenital centers for documenting and improving outcomes, education, and research.
- Each regional ACHD center should establish or affiliate with a patient advocacy group.

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

David J. Skorton, MD, FACC, *Co-Chair*, Arthur Garson, Jr., MD, MPH, MACC, *Co-Chair*, Hugh D. Allen, MD, FACC, James M. Fox, MS, MD, Susie C. Truesdell, PA, MBA, Gary D. Webb, MD, FACC, Roberta G. Williams, MD, FACC

INTRODUCTION

Access to optimal, specialized, appropriate health care, health and life insurance, and full employment remains a problem for many adolescent and adult patients with congenital heart disease (CHD) (1).

Health insurance may be difficult to obtain in adulthood because of "pre-existing conditions"—despite recent federal legislation—and because of uncertainties and misconceptions about the cost of care for adults with CHD. The actual costs of medical care appear to be relatively low in these patients compared with survivors of other chronic diseases

that begin in childhood (2). The costs in these patients, compared with the costs in age-matched patients with adult-onset disease of comparable severity, are unknown.

Federal regulation should provide the opportunity for individuals with disabilities to seek employment, the major source of health insurance coverage for most Americans. The Americans with Disabilities Act does not, however, require that insurance companies change their underwriting approaches or assessment of risk. Companies should not discriminate in hiring on the basis of increased health insurance costs incurred by the disabled.

Patients with CHD may have difficulty obtaining life insurance. Life insurance coverage is now generally more readily available for patients with CHD than it has been in the past. However, life insurance may be unavailable or require elevated premiums for patients with CHD, compared with age-matched control subjects, on the basis of their diagnosis. If financial gain or equity is an issue, alternatives such as savings plans, mutual fund plans, annuity policies, or other, more standard means of investment, may offer similar benefits.

Employment of adults with CHD in an appropriate position and at an appropriate level may require special counseling, for physical and psychosocial reasons. The use of professional job training, vocational rehabilitation, and similar services should be maximized. Recent legislation has attempted to ameliorate this problem for a broad variety of individuals, including those with a wide spectrum of medical disorders. However, full, appropriate employment remains an unfulfilled goal for many adults with CHD.

Organized, effective, and passionate advocacy for adolescents and adults with CHD has been lacking, especially when compared with that of other patients with congenital anomalies and diseases (e.g., National Organization for Rare Disorders, Genetic Alliance). Health care providers and patient groups at local, state, and national levels should intensify efforts to make the needs of these patients more visible and to seek innovative, effective solutions to problems of access.

Access to health care professionals trained appropriately to treat this patient population also remains a challenge. In some academic health centers, special clinics focusing on these patients have been established, but the capacity of these clinics is not adequate to accommodate this growing patient group, as discussed earlier in this Conference report. Access to specialized care in rural areas appears to be a particularly challenging problem.

COST

Data on cost of CHD: a multicenter study. There are relatively few studies defining the life-time costs associated with chronic diseases in children. In 1994, Garson et al. (2) described a multicenter assessment of lifetime costs of care for children with CHD. The study aimed to define total costs associated with the clinical cardiovascular-related care for children with CHD. To accomplish this cost definition, the investigators employed clinical functional categories, with subcategories based on disease severity and treatment options. They also identified six large clinical care sites willing to participate in the collection of cost and clinical practice data, and they extrapolated lifetime costs on the basis of these data. Physicians in each of the six sites assigned typical clinical courses to each subcategory. The clinical outcome was defined by the frequency of seven services: routine clinic visit, complex clinic visit, hospital admission for medical treatment, hospital admission for surgical treatment, hospital admission for interventional treatment, hospital admission for pacemaker implantation, and number of years the patient has taken cardiac medication. Physicians were asked to estimate the percentage of patients who fell into each clinical category and the number of services they would need during the first 40 years of their life. Finally, they were asked to indicate the average charge for each of the services listed.

This study produced the first reliable data on cost and practice variation in pediatric cardiology. Both measures may be used as the basis for increasing control of clinical practice by a variety of influences (e.g., managed care, development of clinical practice guidelines).

Findings: cost and variability. This study provides factual data that can be used to estimate current and future health care costs. Average charges for care (birth to 40 years of age) varied from $47,515 to $73,606, or $650 per year. A simple ratio of charges to mortality was calculated. Although in the early 1990s charges could be used as a surrogate of cost, this is no longer the case. However, the study provides statistics that should be of use to insurers and hospitals in projecting overall cardiovascular costs across a wide range of ages and diagnostic categories. It does not take into account noncardiovascular costs associated with the care of these children. For example, general pediatric care costs incurred by these children were not studied, nor were the costs of respiratory, physical, or occupational therapy and services providing care for children with disabilities, as well as other costs.

Although this study made considerable progress toward identifying cost/benefit ratios based on mortality, the cost/benefit ratios used to determine the validity of new treatment modalities must also include more refined measures of morbidity. The need for more refined definitions of morbidity will enhance the ability to define an optimal outcome. In addition, issues of psychosocial stability, education level, and employability will more adequately define the value of the investment in these children.

Garson et al. (2) also identified substantial variability in practice across institutions. Actual variability in total charges was not as great as practice differences would suggest. However, variability in practice patterns contributes to uncertainty of actual costs.

Some future challenges in assessing costs. YEARLY TREATMENT MODALITY AND OUTCOME VARIABILITY. Estimates of total service utilization in the study of Garson et al. (2)

were based on 1992 utilization. Patients born more recently may have a substantially different outlook from those born in the early 1970s or before. Thus, estimates of total costs of the study are most applicable to the present adult population and have less relevance to infants or children who are currently under care for cyanotic or acyanotic CHD.

SITE OF OUTPATIENT CARE AND INTERPRACTITIONER VARIABILITY. The utilization of services and the frequency of those services may depend on the site of care for adults with CHD. If a pediatric cardiologist cares for them exclusively, their care may be quite different from the care provided by an adult cardiologist, an internist, or an interdisciplinary group focused on adults with CHD.

COSTS BESIDES PHYSICIAN CARE. Only estimated direct medical care costs were included in the study of Garson et al. (2). Two significant costs were excluded: first, the costs to the family—loss of work (i.e., income) for parents, costs of uncovered medical services and drugs, costs of psychologists, and other costs. Second, there are the societal costs associated with loss of work, increased health care needs, and increased educational services. More data on the costs of care are needed. An update of the type of investigation conducted by Garson et al. (2) would be most helpful.

INSURABILITY

After over a decade of efforts to obtain insurance coverage for adults with CHD, some progress has been made, but not enough. Several possible reasons are suggested.

The population. The unique population of young adults with heart disease was projected to include over a million people as we entered the new millennium (3). It was estimated that, after cardiac surgery, 8,500 young adult patients reach adulthood each year (3). Many have chronic, symptomatic cardiac conditions; others are totally asymptomatic, with only mild congenital lesions, such as a small ventricular septal defect. Many have had surgery, some expecting further operations. As operative results and postoperative care continue to improve, the number of young adults with CHD will undoubtedly continue to increase. According to the Second Natural History Study, many patients classified into "simple" diagnostic categories are appropriately educated or employed, or both (4).

Types of insurance. LIFE INSURANCE. Although it is not necessarily considered the best long-term financial investment, life insurance is now considered less of a necessity than it was a few decades ago, because other investment vehicles are available. However, some families consider this a necessary component of their financial planning. Life insurance is now available to more young people with heart defects than it was in the past (1,5,6), but it is still more difficult to obtain for them, compared with individuals with no health problems (7). The implied risk associated with different defects is quite variable among different insurance companies. Some offer standard policies to patients who have mild pulmonary stenosis or closed or small ventricular or atrial septal defects, while others increase the premium rate even for innocent murmurs. They also tend to offer policies more readily to patients who have passed their 15th birthday, assuming that passage from childhood lowers their risk. The cardiologist is often asked to write a letter to the insurance company about the patient's condition. The physician should do so, explaining the long-term expectations regarding the particular patient. In addition, the family should be encouraged to apply for insurance from several companies. Sometimes, using an independent agent will achieve the best results.

HEALTH INSURANCE. Presently, almost 45 million people in the U.S. do not have health care coverage. If the patient's family is fortunate enough to have health insurance, young adults with heart disease can be covered as a dependent until age 19, unless they are still in school or disabled. If more than half-time schooling is pursued, various companies' insurance coverage continues until the patient's 21st or 25th birthday. If the patient's status changes (e.g., by marriage), dependent coverage is often lost. Until age 18 to 21 years, patients may qualify, depending on income levels, for public programs such as Medicaid or State Title V, Children with Special Health Care Needs (CSHCN) Program. The name of these programs varies from state to state. For a directory of such programs, including program name, contact information, eligibility criteria, and scope of services, refer to the Directory of State Title V, CSHCN Programs: Eligibility Criteria and Scope of Services (2000 edition), by John Reiss and Diana Lamar (editors), Gainesville, Florida: Institute for Child Health Policy (http://www.ichp.edu). Others who qualify for Social Security (by virtue of being determined to be disabled) can obtain Medicare, Part A coverage, but must purchase Medicare, Part B for 80% physician services (20% co-pay).

Previous studies have indicated that between 10% and 22% of adults with CHD are uninsured, and 67% have reported difficulty in obtaining health insurance or changing jobs to guarantee coverage (8). Those with a history of surgical repair reported the greatest difficulty, although this may not correlate with their current severity of illness. Most commonly, patients can obtain insurance only after the exclusion of cardiac disease as a pre-existing illness, by paying higher premiums to participate in a high-risk reinsurance pool, or by obtaining coverage through their employer, in either a health maintenance organization or self-insured plan.

Types of coverage vary. The common type of coverage 10 to 20 years ago was an independent health care policy. Now most people have some form of group coverage, usually purchased through their employer. Most of these plans are "managed" (i.e., they are linked to a network of participating physicians and hospitals). In the most developed health maintenance organization, the choice of physicians is usually restricted to the network, and a primary care physician ("gatekeeper") usually directs care by a specialist (e.g.,

cardiologist). The gatekeeper may be directed by an internally developed or nationally developed set of guidelines for specialty care referral. In the most rigid circumstances, the gatekeeper may be the health maintenance organization itself, which may actually direct the referral to the cardiologist of its choice. In a point-of-service plan or a preferred provider organization, patients can go freely out of the network to choose a specialist, with a plan-defined deductible and co-payment, representing a greater financial responsibility for the patient, compared with obtaining care within the network. Within this organizational framework, it may be difficult for adolescents or adults with CHD to access care by a skilled cardiologist who is either familiar with or has expertise in CHD. This can lead to underutilization (withholding of specialty access or testing) or over-utilization (unnecessary testing performed by a cardiologist inexperienced in caring for adults with CHD).

There is a similar obstacle to the team concept that is crucial in the care of the adolescent and adult patient with CHD and associated or other health problems. Referral and reimbursement to multiple subspecialists and mid-level provider team members may be a new concept for the insurer, who may not understand and who may reject this option for the patient. There is a clear need to educate the insurer about this care delivery model, compared with the multidisciplinary model that is accepted by most insurers for the diagnosis and treatment of cancer.

In summary, the current managed care insurance model does not easily support what may be considered the optimal care of adults with CHD. There needs to be a recognition by insurers that the complex range of conditions and the care needs of adults with CHD are different from those of adults with acquired heart disease, and current referral and reimbursement models are inadequate to address these differences.

What can be done? In the early 1990s, the Council on Cardiovascular Disease in the Young, of the American Heart Association (AHA), held a conference on insurability of young adults with heart disease (3). Suggestions from that conference generally apply today and can still be considered.

As practice guidelines relevant to adults with CHD are developed, organizations such as the American College of Cardiology (ACC) should share and discuss these guidelines with insurers. Such guidelines should allow insurers to more accurately project costs of care and to develop appropriate premiums.

It was also suggested at the AHA conference (3) that companies use community standards in the development of premiums, rather than using small-group standards or a claims-made standard that penalizes a given patient or family. The use of clinical practice guidelines should encourage the appropriate use of technology, thus further controlling costs. Training programs should develop strategies to produce a cadre of adequately trained cardiologists who can provide optimal and cost-effective care to this population. Uniform coding and billing processes would greatly reduce paperwork and office/hospital overhead.

Preventive care for pregnant women, which is often not available to the poor (9), could reduce some cases of CHD in newborns, as caused by the mother's alcohol and cocaine abuse, as well as infections that could have been prevented by immunization. Prevention of premature birth would also improve survival and decrease the incidence of some childhood diseases. Genetic counseling is also important for couples whose offspring are at increased risk of CHD.

Over the past two decades, patients have sometimes had to resort to drastic measures (e.g., quitting work, remaining in an unsatisfactory position) to maintain health insurance coverage. Unfortunately, some have elected to avoid clinic visits, catheterization, or operation because of the personal financial consequences. Some have died as a result; many have suffered an unnecessary decline in function. None of these choices is appropriate or fair.

Since State Title V CSHCN programs cover cystic fibrosis and hemophilia after the age of 21 years, why not do so for CHD? This is an avenue that should be pursued.

EMPLOYABILITY AND VOCATIONAL SUPPORT

Employment status. Reports of employment status of adults with CHD vary. No more than 10% are considered totally disabled. Those with a mild disability reported a 50% increased rejection rate in job applications, and those with a moderate-to-severe disability reported a 400% increase in rejections of job applications, in comparison with nondisabled control subjects. The severity of disability has been correlated with unemployment and lower income (10). There have been numerous assessments of employment status of adults with CHD in the last decade (10,11), with 8% to 13% receiving public assistance or living as a dependent with relatives.

U.S. federal regulations. Existing federal regulations provide for training and improved prospects for employment of people considered disabled (e.g., Vocational Rehabilitation Act). Subsequently, there have been further congressional acts barring employment discrimination by any federal employer or employer receiving federal funding (Rehabilitation Act of 1973); the U.S. Civil Service (Act of June 10, 1948) and the Americans with Disabilities Act (ADA of 1990), which extends this provision to the private sector, are two other such acts. Most recently, the Work Incentives Improvement Act was passed in 1999; this act provides for a stepped approach to less severely disabled individuals who could reasonably be expected to be functional and employable with assistance.

The Rehabilitation Act of 1973 also established affirmative action for the advancement of disabled persons, including hiring, placement, and vocational rehabilitation. It also provided for the National Council on the Handicapped to be formed within the Department of Health, Education and Welfare. This council was later granted the authority to

review all federal laws and programs regarding individuals with disabilities.

The ADA prohibits discrimination with respect to hiring, promotion, or discharge of employees on the basis of disability. Employers are also required to make accommodations, within reason, to allow a disabled employee to perform a job. Although the ADA specifically excludes insurance coverage practices from these injunctions, employers cannot deny employment on the basis of the coverage, or lack of coverage, provided by their insurance benefits, or because their cost of insurance would increase.

The Work Incentives Improvement Act allows for state-sponsored Medicaid programs to cover some adults who may be declared "disabled" by virtue of their underlying condition. The legislation allows states to define the list of conditions. Therefore, it is possible that a state could define adults with CHD as "disabled" and eligible for coverage; this would require each ACC chapter to work with state Medicaid programs and state legislators to define the eligibility. The ACC Advocacy Division has resources for chapters to help in this effort.

Strategies to assist in employment counseling. The most important element in employment counseling by the health care provider is an expert, realistic, and assertive estimate of the patient's physical capabilities as they relate to available vocational options. Once this is done, services such as vocational rehabilitation, job training, and physical rehabilitation can be offered. The practitioner should also strongly consider direct involvement with the employer, at the patient's request, to assist in an optimal match between patient capabilities and job requirements. Despite shortfalls in legislation and health care coverage, concerted efforts made by the health care provider can make an enormous difference in a specific patient's vocational experience.

ADVOCACY

The ACC has made a strong statement supporting access to cardiovascular care, regardless of a patient's ability to pay (12). Recently, a plan to achieve universal coverage by 2010 was the topic of the ACC Presidential Plenary Address; this has been published in the *Journal of the American College of Cardiology* (13). In the meantime, however, we have today's reality and must take incremental steps to provide coverage for this segment of the population.

Most of these patients are not severely disabled and are capable of working and contributing to society (8,14). Despite this, insurance coverage is denied, limited, or associated with unacceptably high premiums. For example, through state high-risk pools, premiums for these individuals may exceed standard premiums by as much as 50%, making this form of insurance inaccessible for many of the people who need it most. Indeed, as indicated previously, even those who have insurance face other issues, such as under-insurance, disapproval by managed care companies of specific medical services, and life-time caps on coverage.

This population of individuals is particularly vulnerable because they suffer from conditions they have had all or most of their lives. They have received coverage and treatment as children, only to have it taken away at a time in their lives when they are expected to become self-sufficient. To complicate matters, these patients, who are generally capable of working, often have difficulty finding employment because of their health history.

The ACC's legislative approach. The ACC leadership has met with staff at the White House, members of Congress, and numerous other specialty and patient organizations to discuss possible mechanisms for providing health insurance and job training to those with childhood diseases, including CHD. The ACC presented a resolution to the American Medical Association (AMA) House of Delegates; the AMA issued a report in December 1999 (15) encouraging the government to identify these individuals and the barriers to their care.

The ACC worked on a popular proposal introduced by Senator Edward M. Kennedy, D-Mass.; Senator James Jeffords, R-Vt.; and Rep. Rick Lazio, R-N.Y. Endorsed by the Administration, this legislation—the Work Incentives Improvement Act described earlier—provides an incremental approach to addressing the health insurance needs of the less severely disabled. The overriding intent of the legislation is to enable disabled individuals to return to work, but it also contains a provision that allows state demonstration projects for people who are less severely disabled and who, in the absence of needed health care services, would reasonably be expected to become disabled. The demonstrations specifically permit states to offer these individuals a Medicaid buy-in option. The ACC worked with legislators to add to the House Commerce Committee report accompanying the bill, language that clarifies the congressional intent of the proposal. The report's new language says that states could include in the definition of "potentially severe disability," those individuals with congenital birth defects or other diseases developed in childhood. The ACC key contacts were alerted, and they provided important support. The bill was signed into law in 1999. This is an important first step.

The role of ACC chapters. Individual ACC chapters are encouraged to take up this issue on behalf of patients with cardiac diseases. Already, individuals in some states are considering proposals to fulfill the impending mandates of the Work Incentives Improvement Act. The ACC has materials ready to assist chapters in assessing the scope of the problem in individual states and determining strategies for communicating with state officials.

The role of individual physicians. Our patients need us to advise them about what to expect in the real world. We should tell our patients before they enter adulthood that their health insurance coverage requires their attention and should be of concern; they should be advised to seek jobs, as appropriate, with large employers or the state or federal government. Our patients should understand that, under

law, their health status is to have no bearing on employment; therefore, employers are generally not permitted to inquire about their condition. This advice can help the patients we know, but we must also endeavor to help those we do not know, by working with our legislators to extend coverage to as many people as possible.

RECOMMENDATIONS

Based on the considerations outlined earlier, the Task Force recommends that the ACC take the following actions:

General

- Develop a strategic plan for organized advocacy for this patient population to include health care professionals, patients, and their families, in the context of a public relations campaign.

Insurance Coverage and Health Care Costs

- Develop educational materials to guide adolescent and adult patients in the transition to independence, including the need for health (and perhaps life) insurance, barriers that may exist in obtaining coverage, and strategies to obtain optimal coverage.
- Develop a better understanding of the true economic impact (e.g., payments, future income potential) of CHD in adults; this will involve sponsoring a multicenter study with economic forecasting.
- Include, in formal and regular discussions with insurance companies and other public and private payors and purchasers, information on the special problems encountered and expertise necessary in the care of adolescents and adults with CHD.
- Reduce the barriers to multidisciplinary services by developing innovative reimbursement methodologies. Pilot programs established between one or more ACHD centers and major payors (public and private) should be encouraged.
- Work, at the chapter level, with state legislators to specify CHD in a demonstration project of the Work Incentives Improvement Act.
- Recommend that physicians discuss individual patient coverage concerns with insurance company medical directors.
- Advocate health care coverage for all. As an incremental step, all adults with CHD should be covered, thus removing a significant barrier to access.

Education, Employability, and Vocational Counseling

- Develop additional educational materials to help adolescent and adult patients as they approach the job market, focusing on their legal rights (e.g., health should not be discussed during an interview), tips for success, and where to go for job training and vocational counseling.
- Recommend that, at the patient's request, individual physicians work directly with patients, their schools, and their employers or potential employers to optimize opportunities.

REFERENCES

1. Mahoney LT, Skorton DJ. Insurability and employability. J Am Coll Cardiol 1991;18:334–6.
2. Garson A, Jr., Allen HD, Gersony WM, et al. The cost of congenital heart disease in children and adults: a model for multicenter assessment of price and practice variation. Arch Pediatr Adolesc Med 1994;148:1039–45.
3. Allen HD, Gersony WM, Taubert KA. Insurability of the adolescent and young adult with heart disease: report from the Fifth Conference on Insurability, October 3–4, 1991, Columbus, Ohio. Circulation 1992;86:703–10.
4. Gersony WM, Hayes CJ, Driscoll DJ, et al. Second natural history study of congenital heart defects: quality of life of patients with aortic stenosis, pulmonary stenosis, or ventricular septal defect. Circulation 1993;87 Suppl I:I-52–65.
5. Manning J. Insurability and employability of young cardiac patients. In: Engle M, editor. Pediatric Cardiovascular Disease. Philadelphia: Davis, 1981:117–27.
6. Ad Hoc Committee of the Council on Cardiovascular Disease in the Young. AHA Committee report: guidelines for insurability of patients with congenital heart disease. Based on the Fourth Conference on Insurability of Young Cardiacs, October, 1977, with revisions and updating June 1979. Circulation 1980;62:1419A–24A.
7. Truesdell SC, Skorton DJ, Lauer RM. Life insurance for children with cardiovascular disease. Pediatrics 1986;77:687–91.
8. Truesdell SC, Clark EB. Health insurance status in a cohort of children and young adults with congenital cardiac diagnoses (abstract). Circulation 1991;84 Suppl II:II-386.
9. Allen HD, Taubert KA, Deckelbaum RJ, et al. Poverty and cardiac disease in children. Am J Dis Child 1991;145:550–3.
10. Celermajer DS, Deanfield JE. Employment and insurance for young adults with congenital heart disease. Br Heart J 1993;69:539–43.
11. McGrath KA, Truesdell SC. Employability and career counseling for adolescents and adults with congenital heart disease. Nurs Clin North Am 1994;29:319–30.
12. Hutter AM, Jr., Alpert JS, Cheitlin MD, et al. Access to cardiovascular care and related issues: Ad Hoc Task Force on Access to Cardiovascular Care, American College of Cardiology. J Am Coll Cardiol 1998;31:485–6.
13. Garson A, Jr. The U.S. health care system 2010: problems, principles, and potential solutions (President's page). J Am Coll Cardiol 2000; 35:1048–52.
14. Garson A, Jr. Health care policy for adults with congenital heart disease: the patient, the physician, and society. Circulation 1992;86: 1030–2.
15. American Medical Association Council on Medical Services (CMS). Status report on increasing access for the uninsured. CMS Report 2, A-99, 1999.