Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues: A Scientific Statement From the American Heart Association

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Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues

A Scientific Statement From the American Heart Association

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Many children born with complex childhood illnesses that historically caused early death are now surviving into adulthood with the expectation of leading meaningful and productive lives. They will ultimately need to transition their care from pediatric to adult-centered care. Unfortunately, in the absence of structured programs to guide this transition, there is often delayed or inappropriate care, improper timing of the transfer of care, and undue emotional and financial stress on the patients, their families, and the healthcare system. At its worst, and as frequently happens now, patients are lost to appropriate follow-up. In fact, the number of adults with congenital heart disease (CHD) in the United States is rising exponentially and now exceeds 1,000,000.1–7 At least half of these patients may have complex CHD. Fewer than 30% of adults with CHD are seen by appropriate specialized providers. Fewer than 15% of these patients, who are seen in specialty adult CHD (ACHD) clinics, have CHD that is classified as severe.8 Thus, adolescents with CHD constitute a growing population of individuals for whom a well-planned and well-executed “transition process” is essential.

The goals of a formal transition program are to prepare young adults for transfer of care. It should provide uninterrupted health care that is patient centered, age and developmentally appropriate, flexible, and comprehensive. It should include age-appropriate education about medical conditions and promote skills in communication, decision making, self-care, and self-advocacy.9–13 It should foster greater personal and medical independence and a greater sense of control over health, healthcare decisions, and psychosocial environment. The ultimate goal of a transition program is to optimize the quality of life (QOL), life expectancy, and future productivity of young patients.14

We acknowledge that the development of ideal transition programs is a laudable goal that may not be achievable in the current healthcare environment. With the recognition that...
adolescents and young adults receive their care from a diverse group of providers, this document is intended for pediatric and adult cardiologists who are likely to care for this group of patients. We have addressed diverse topics that pertain to the care of adolescents and young adults with CHD, beginning with issues directly related to transition. The American Heart Association (AHA) classification of recommendations and levels of evidence for practice guidelines were used when applicable. The classification and levels of evidence are shown in Table 1.

Concepts of Transition Care
In the United States, ~14% of children are identified as having a special healthcare need. More than 1 in every 5 households has at least 1 child with some special needs. This number represents a slight increase from data reported in 2001, perhaps because of increasing diagnostic capability, increased medical access, and an increasing survival rate for many congenital or chronic conditions that historically were associated with high childhood mortality. Advances in medical and surgical care for children with congenital cardiovascular disease have dramatically improved their life expectancy, resulting in 85% to 90% of these children surviving to adulthood.

Transition is defined by the Merriam-Webster dictionary as “a movement, development, or evolution from one form, stage, or style to another.” Medical transition is the process of moving from a pediatric medical system to an adult one. Transfer refers to the actual point in time at which responsibility for patient care is “handed off” to the adult provider. Developmental transitions occur at specific milestones throughout the life spectrum.

The American Academy of Pediatrics states, “The goal of transition in health care for young adults with special health care needs is to maximize lifelong functioning and potential through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood.” However, for children with congenital or long-term health-care needs, transition to an adult lifestyle or environment is often made more difficult by the inability to obtain quality health care as they leave their pediatric providers. The American Academy of Pediatrics’ Medical Home Initiative states that 90% of children with special needs reach their 21st birthday, yet 45% of them lack access to a physician who is familiar with their condition(s). In addition, 30% of all young adults 18 to 24 years of age lack a payment source for their health care. The Maternal Child Health Bureau, a division of the Health Resources and Services Administration, US Department of Health and Human Services, in response to Healthy People 2010 national health objectives and the President’s New Freedom Initiative, includes “transition to adult health care, work, and independence” in 6 core systems changes that are deemed necessary to improve the overall health of children with special needs.

To date, transition outcomes overall have been shown to be suboptimal, particularly among ethnic minorities. Better outcomes are seen for older teens and those receiving care within a “medical home.” Several steps have been identified that are paramount to the successful transition of a child with special needs from a family-centered pediatric system to a patient-centered adult long-term care system, including the following:

1. Comprehensive care that is coordinated and managed through a medical home (usually a primary care provider but in some instances a tertiary care center or a subspecialty practice)
2. Access to healthcare financing
3. Education of adult providers in managing chronic conditions previously limited to the pediatric population
4. Ongoing, coordinated communication between patients, families, and pediatric and adult healthcare providers to facilitate transition and transfer

Reid et al21 reported in 2004 that only 48% of adolescents with CHD underwent successful transition. Their data suggested that ongoing care during adolescence with continued discussion of the importance of transition was very important to successful transition. The standard of transition as defined by the Society of Adolescent Medicine is “a purposeful, planned process that addresses the medical, psychosocial and educational/vocational needs of adolescents and young adults with chronic physical and medical conditions as they move from child-centered to adult-oriented health care systems.” The aim of the present statement is to provide guidelines for clinicians to promote early and high-quality care and support for their patients with CHD for successful transition of care.

Timing of Transition
The transitional process depends on the patient’s medical and developmental status and should be individualized.22 The developmental status of the patient should be considered relative to normal developmental milestones (Table 2), with special considerations for how chronic illness (Table 3) and congenital heart defects (Table 4) impact these milestones. The American College of Cardiology task force and the recent “ACC/AHA Guidelines on the Management of Adults With Congenital Heart Disease” recommended that the transition process start at 12 years of age to prepare the patient for transfer to adult care.

“Envisioning a future” is an important first stage in the transition process and occurs at the time of diagnosis of a chronic condition.24 This emphasizes starting the discussion early and includes prompting parents to consider future expectations for their child’s education, employment, and independent living.24 Just as patients are prepared to take increased responsibility for their health care, parents also benefit from education and support as they gradually share and then hand over the primary responsibility for healthcare management. From initial diagnosis, discussion with parents should reference transitional topics to help parents prepare for this shift in responsibility. Preparing the patient for self-care is an ongoing process that should begin in early childhood and continue into adulthood to allow the development of the necessary self-care skills.26 Knowledge of the developmental stages in adolescence and the impact of chronic illness is essential for a successful transition.27 During childhood, the patient should become increasingly involved in direct discussions about his or her diagnosis, medications, and exercise...
limitations. In the teenage years, discussions about heart-healthy behaviors and the risks of smoking, alcohol, and drugs should be introduced and reviewed regularly. Vocational and employment advice should continue into early adulthood, and birth control, pregnancy, genetics, and long-term prognosis should also be emphasized. Table 3 describes practical steps that can be undertaken in the designated age group, with the assumption of appropriate developmental status. In patients with developmental delay, this timeline will have to be altered as appropriate. Self-care education should not culminate with transfer to adult care but is an ongoing educational process to address life changes. Even patients who continue to see the same healthcare providers for CHD care should undergo a process of transition similar to that provided by family practitioners and internal medicine/pediatric specialists who do not transfer patient care.

Before transfer to ACHD care, ensure that any ongoing medical or surgical issues are addressed. A portable and accessible summary of medical history allows for effective collaboration among healthcare providers. A transfer-of-care letter with relevant medical and psychosocial information and a care plan should be provided to the regional ACHD center in a timely fashion, with a copy for the patient to keep in case the ACHD provider changes.

The simultaneous transfer of care of the young adult to adult general care and subspecialty care during a period of significant life changes can be overwhelming. This simulta-

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Table 1. Applying Classification of Recommendation and Level of Evidence

<table>
<thead>
<tr>
<th>SIZE OF TREATMENT EFFECT</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CLASS I</strong> Benefit &gt;&gt; Risk</td>
</tr>
<tr>
<td>Procedure/Treatment SHOULD be performed/ administered</td>
</tr>
</tbody>
</table>

| **CLASS IIa** Benefit > Risk |
| Additional studies with focused objectives needed |
| IT IS REASONABLE to perform procedure/administer treatment |

| **CLASS IIb** Benefit ≥ Risk |
| Additional studies with broad objectives needed; additional registry data would be helpful |
| Procedure/Treatment MAY BE CONSIDERED |

| **CLASS III** No Benefit or CLASS III Harm |
| Procedure/ Test |
| Treatment |
| COR III: No Benefit |
| Not Helpful |
| No Proven Benefit |

**LEVEL A**
- Recommendation that procedure or treatment is useful/effective
- Sufficient evidence from multiple randomized trials or meta-analyses

**LEVEL B**
- Recommendation that procedure or treatment is useful/effective
- Evidence from single randomized trial or nonrandomized studies

**LEVEL C**
- Recommendation that procedure or treatment is useful/effective
- Only expert opinion, case studies, or standard of care

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*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as sex, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Although randomized trials are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

†For comparative effectiveness recommendations (Class I and IIa; Level of Evidence A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.
Table 2. Typical Life Span Developmental Tasks After Childhood for Individuals Without a Chronic Illness,4 by Age Group

<table>
<thead>
<tr>
<th>Domains</th>
<th>Mid Adolescence (14–16 y)</th>
<th>Late Adolescence (16–19 y)</th>
<th>Young Adulthood (19–35 y)</th>
<th>Middle Adulthood (Mid 30s–Mid 40s)</th>
<th>Maturity (Mid 40s–Mid 60s)</th>
<th>Old Age (Mid 60s+)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical</td>
<td>Completing puberty Positive body image</td>
<td>Maintaining physical health Positive body image</td>
<td>Maintaining physical health</td>
<td>Accepting physiological changes</td>
<td>Accepting physiological changes and onset of common illnesses of adulthood</td>
<td>Adjusting to decreased physical health and/or illness</td>
</tr>
<tr>
<td>Social and family relations</td>
<td>Dating Establishing peer relationships</td>
<td>Separation from family of origin Selecting a mate/partner Starting a family Finding a social group</td>
<td>Childrearing</td>
<td>Assisting teenage children in developing independence Adjusting to aging parents</td>
<td>Grandparenting Coping with loss of peers</td>
<td></td>
</tr>
<tr>
<td>Emotional</td>
<td>Competence in emotional self-regulation</td>
<td>Competence in emotional self-regulation</td>
<td>Maintaining emotional/ mental health</td>
<td>Maintaining emotional/ mental health</td>
<td>Maintaining emotional/ mental health</td>
<td>Maintaining emotional/ mental health</td>
</tr>
<tr>
<td>Education and vocation</td>
<td>Completing high school Higher education and/or entering work force</td>
<td>Establishing and/or maintaining an occupation</td>
<td>Maintaining standard of living</td>
<td>Satisfaction with occupational productivity</td>
<td>Retirement</td>
<td></td>
</tr>
<tr>
<td>Health behaviors</td>
<td>Appropriate exercise and diet Avoiding health-risk behaviors Maintaining appropriate weight</td>
<td>Appropriate exercise and diet Avoiding health-risk behaviors Maintaining appropriate weight</td>
<td>Appropriate exercise and diet Maintaining appropriate weight Obtaining screening for health problems as recommended for age and sex</td>
<td>Appropriate exercise and diet Maintaining appropriate weight Obtaining screening for health problems as recommended for age and sex</td>
<td>Appropriate exercise and diet Maintaining appropriate weight Obtaining screening for health problems as recommended for age and sex</td>
<td></td>
</tr>
<tr>
<td>Personality and identity</td>
<td>Sex-role development Increasing independence</td>
<td>Balancing independence and interdependence with family and friends</td>
<td>Balancing independence and interdependence with family and friends</td>
<td>Balancing independence and interdependence with family and friends</td>
<td>Acceptance of one’s life history Acceptance of death</td>
<td></td>
</tr>
</tbody>
</table>

Neuropsychological development can be avoided by transitioning the adolescent with chronic health needs to a family practice or an internal medicine/pediatric provider or to a medical care home for primary care. This provider can then follow the patient into adulthood while the young adult is transferred to ACHD care. Consider referring patients with complex medical conditions to a regional “medical home,”29 because that facilitates better medical transition.20

Social/Family Dynamics: Impact of Chronic Illness as Related to CHD

Patients

CHD is a chronic illness, and as such, the need for long-term medical monitoring, medication adjustments, repeated hospitalizations, and illness-related absences from school can affect the adolescent’s self-image and self-esteem, as well as delay completion of normal developmental tasks.27,30 During the transition process, it is important for providers to understand how the individual adolescent and his or her family interpret the diagnosis and prognosis. Research consistently has shown that psychological adjustment is not proportionally related to the severity of the cardiac defect.31–36 Healthcare providers must also take care not to place their own judgment value on an adolescent’s subjective experience and coping process. Cultural and family dynamics and past experiences both within and outside of the healthcare environment are other important factors to consider when preparing adolescents and their families for the transition process.

Early research reported conflicting results about the emotional and behavioral problems in adolescents with CHD. Although some studies reported anxiety, diminished self-esteem, depression, increased dependency, and poor emotional and social adjustment among adolescents with CHD,37–40 others reported no significant psychopathology.41,42 This discrepancy has been attributed to differences in methodology, types of defects, and small sample size.43 A recent meta-analysis of 11 studies of the psychological problems of children and adolescents with CHD revealed that older children and adolescents had an increased risk of internalizing problems (eg, depression, anxiety) and externalizing behavior problems (eg, aggression, hyperactivity) compared with healthy control subjects.44 One study examined patients at a single institution before 1980 and a decade later to ascertain whether newer treatment modalities and improved surgical outcomes provided more favorable psychosocial outcomes. No differences were found; improvements in psychological outcomes did not track with medical outcomes.44 On balance, the percentage of adolescents and adults with CHD who have emotional and behavioral problems is slightly greater than in the general population.45

Qualitative research using semistructured interviews has recently provided insight into the social experiences of adolescents with CHD. Studies suggest that although adolescents with CHD may try to live a normal life, these teens describe feeling “different” and set apart from their peers because of their heart disease.46 Adolescents speak of com-
Table 3. Transition Timeline for Adolescents With Special Healthcare Needs: Chronic Illnesses/Physical Disabilities

<table>
<thead>
<tr>
<th>Birth to 3–5 y, or according to your child’s developmental ability</th>
<th>By ages 6–11 y, or according to your child’s developmental ability</th>
<th>By ages 12–18 y, or according to your child’s developmental ability</th>
<th>By ages 18–21 y, or according to your child’s developmental ability</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Begin keeping a record of your child’s early intervention, educational, and medical history, including immunizations.</td>
<td>• Assess your child’s perception and basic knowledge of his/her heart condition. Build on their understanding.</td>
<td>• Assess your teen’s perception and basic knowledge of his/her heart condition. Fill in gaps in understanding.</td>
<td>• Act as a resource and support to your young adult.</td>
</tr>
<tr>
<td>• Attend support groups and get to know other parents who have children with a chronic illness. Learn from their experiences, be encouraged, and find out about helpful resources.</td>
<td>• Continue teaching your child general self-care and health skills, as well as skills related to his/her special healthcare need.</td>
<td>• Continue teaching your teen general self-help and health skills, as well as skills related to his/her special healthcare need.</td>
<td>• Encourage your young adult to participate in support groups and/or organizations relevant to his/her special healthcare need.</td>
</tr>
<tr>
<td>• By ages 3–5 y, or according to your child’s developmental ability</td>
<td>• Discuss relationships and personal safety with your child.</td>
<td>• Begin helping your teen keep a record of his/her medical history, including conditions, operations, treatments (dates, doctors, recommendations) and 504 plan if he/she has one.</td>
<td>• Finalize healthcare coverage with your young adult.</td>
</tr>
<tr>
<td>• Assign your child chores appropriate for his/her ability level.</td>
<td>• Determine whether reasonable accommodations are needed to ensure equal access to school programs; if so, ask if your child qualifies for a 504 plan.</td>
<td>• Begin helping your teen find work and volunteer activities.</td>
<td>• With your young adult, finalize transfer of medical care to an adult provider.</td>
</tr>
<tr>
<td>• Encourage decision-making skills by offering choices.</td>
<td>• Encourage hobbies and leisure activities; include exploring community and recreational activities, clubs, 4-H, Scouts, Campfire, YMCA, sports, and so on.</td>
<td>• Discuss relationships, sexuality and personal safety with your teen.</td>
<td>• If your young adult is attending college, encourage continued contact with disabled student services as needed for accommodations.</td>
</tr>
<tr>
<td>• Teach natural consequences of your child’s behaviors and choices.</td>
<td>• Continue to encourage decision-making skills by offering choices.</td>
<td>• Continue teaching your teen general self-advocacy skills.</td>
<td>• Encourage your young adult to investigate services provided by the Department of Vocational Rehabilitation if he/she has not already done so.</td>
</tr>
<tr>
<td>• Continue involvement in community and recreational activities that include children with and without special needs.</td>
<td>• Continue assigning your child chores appropriate for his/her ability level.</td>
<td>• Begin asking “What do you want to do when you grow up?”</td>
<td>• By ages 18–21 y, or according to your child’s developmental ability</td>
</tr>
<tr>
<td>• Begin teaching your child self-care skills: general skills and those related to his/her special healthcare need.</td>
<td>• Take your child shopping whenever possible so he/she can help in choices.</td>
<td>• Let your child choose how to spend some or all of his/her allowance.</td>
<td>• SSI indicates Supplemental Security Income.</td>
</tr>
<tr>
<td>• Begin helping your child interact directly with doctors, nurses, therapists, and teachers.</td>
<td>• Teach your child the consequences of his/her behaviors and choices.</td>
<td>• Teach your child the consequences of a poor choice as well as a good choice.</td>
<td></td>
</tr>
<tr>
<td>• Begin teaching child about relationships, personal space, and their body (including their heart condition).</td>
<td>• Allow your child to experience the consequences of his/her behaviors and choices.</td>
<td>• Begin teaching your child self-advocacy skills.</td>
<td>• Begin teaching your child the consequences of his/her behaviors and choices.</td>
</tr>
</tbody>
</table>

mon dilemmas and concerns, including whether or not to disclose their heart disease to others; the fear of rejection and other effects of disclosure on social relationships; and the fear of forming close relationships. Adolescents may have recognized the need to take a more active role in managing their health care, but they were unsure how to begin. In the report by Tong et al., adolescents were concerned about planning for the future given their uncertain longevity. In another study, both adolescents and young adults tended to have unrealistic expectations of their life expectancy, and qualitative data suggested a lack of awareness and understanding of their future health risks. Compared with their healthy peers, adolescents with CHD are less likely to smoke, drink alcohol, or use illicit drugs and are less likely to be sexually active.
Table 4. Life Span Developmental Tasks and Issues for the Adolescent and Adult With Congenital Heart Defects,* by Age Group

<table>
<thead>
<tr>
<th>Domains</th>
<th>Mid Adolescence (14–16 y)</th>
<th>Late Adolescence (16–19 y)</th>
<th>Young Adulthood (19–35 y)</th>
<th>Middle Adulthood (Mid 30s–Mid 40s)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical</td>
<td>Coping with body image and limitations in physical functioning</td>
<td>Coping with body image and limitations in physical functioning</td>
<td>Gradual or acute decreases in physical functioning; burden/complications with onset of common illnesses of adulthood</td>
<td>Gradual or acute decreases in physical functioning; burden/complications with onset of common illnesses of adulthood</td>
</tr>
<tr>
<td>Social and family relations</td>
<td>Peer acceptance of physical appearance/limitations; coping with stigmatization; lack of social support for CHD issues</td>
<td>Decisions about dating; increasing independence from family; lack of social support for CHD issues</td>
<td>Decisions regarding life partner and reproduction; coping with loss of normative family life cycle; finding a social group/network</td>
<td>Addressing the impact of premature death on partner, any children, and extended family</td>
</tr>
<tr>
<td>Emotional</td>
<td>Managing anxiety-provoking medical procedures; maintaining emotional adjustment during period of critical transitions</td>
<td>Managing anxiety-provoking medical procedures; maintaining emotional adjustment during period of critical transitions</td>
<td>Managing anxiety-provoking medical procedures; avoiding arrhythmia-related anxiety/ phobic reactions; avoiding despair, depression, or anxiety; maintaining emotional/mental health</td>
<td>Managing anxiety-provoking medical procedures; avoiding arrhythmia-related anxiety/ phobic reactions; avoiding despair, depression, or anxiety; maintaining emotional/mental health</td>
</tr>
<tr>
<td>Education and vocation</td>
<td>Coping with possible intellectual and/or learning disabilities</td>
<td>Selecting educational and vocational goals appropriate to present/future abilities</td>
<td>Stigmatization/discrimination in obtaining employment; maintaining employment during medical crises</td>
<td>Maintaining/changing employment and/or career goals with decreases in physical functioning</td>
</tr>
<tr>
<td>Medical</td>
<td>Taking some responsibility for medical care; learning appropriate health behaviors</td>
<td>Increasing responsibility for medical care; transition to adult care; knowledge of diagnosis, prognosis, and associated health behaviors</td>
<td>Primary responsibility for medical care; knowledge of prognosis; reoperation(s); CHD complications; coping with medical procedures and hospitalization; coping with procedure-related pain</td>
<td>Primary responsibility for medical care; knowledge of prognosis; reoperation(s); CHD complications; coping with medical procedures and hospitalization; coping with procedure-related pain</td>
</tr>
<tr>
<td>Health behaviors</td>
<td>Avoiding initiation of risky health behaviors; maintaining appropriate weight and getting exercise; maintaining oral hygiene and preventing endocarditis</td>
<td>Regular medical follow-up; avoiding risky health behaviors; maintaining appropriate weight; getting appropriate exercise; maintaining oral hygiene and preventing endocarditis</td>
<td>Regular medical follow-up; avoiding risky health behaviors; maintaining appropriate weight; getting appropriate exercise; maintaining oral hygiene and preventing endocarditis</td>
<td>Regular medical follow-up; avoiding risky health behaviors; maintaining appropriate weight; getting appropriate exercise; maintaining oral hygiene and preventing endocarditis</td>
</tr>
<tr>
<td>Personality and identity</td>
<td>Integration of CHD into self; acceptance of being different and unique</td>
<td>Lack of control over health outcomes; increasing independence</td>
<td>Balancing independence and interdependence with family and friends</td>
<td>Resolving loss of typical life achievements; facing prospect of premature death</td>
</tr>
</tbody>
</table>

CHD indicates congenital heart disease.

*Life expectancy varies with lesion severity and increases with improved medical care.

Birks et al560 found that although the participants in their study coped well, concerns related to being unable to physically keep up with their peers or consume alcohol as freely as their peers (because of medication side effects) were especially challenging and isolating. Not engaging in common risky behaviors during adolescence is appropriate given the negative health implications; however, this creates additional challenges for social acceptance and integration for adolescents with CHD. Adolescents in another qualitative study developed strategies to deal with the physical limitations and decreased stamina imposed by the disease, but the discrimination and bullying they experienced from peers left them feeling particularly angry and excluded.561

Evaluation of QOL in adolescents with CHD by use of the Pediatric Quality of Life Inventory revealed significantly lower overall perceived QOL, including physical, psychosocial, and school functioning, with the greatest difference in the domain of school functioning. Specific psychosocial issues included feeling angry and being worried about the future, and these were unrelated to the severity of the cardiac defect. In contrast, Culbert et al52 used the Child Health Questionnaire to examine health status or QOL in adolescents with transposition of the great arteries 11 to 15 years after their initial repair and concluded that health status as perceived by this specific cohort was excellent and showed no significant difference from the published normative data. In addition, a more positive outcome was found for subjects who had the arterial switch procedure versus the atrial (Senning/ Mustard) repair.52 Although patients undergoing atrial switch may have had longer periods of cyanosis, they were born
during the same time period as the arterial switch patients (1985–1989).

Overall, the combined quantitative and qualitative data on the QOL of teenagers with CHD suggest that they face unique challenges in addition to those typically confronted during adolescence. Clinically, it is important to understand the reasons behind adolescent perceptions of their health status and QOL to provide appropriate education and support during the transition process.

Parent/Family

Transition planning for adolescents with CHD cannot occur without a tandem evaluation of the impact of the diagnosis on their parents. During their formative years, children and adolescents learn and take cues from their parents on how to manage and adapt to their illness. Although parents may be expected to take an active role in the transition process, some parents find this difficult. Because of the chronic nature of CHD, many parents have devoted substantial time and energy to monitoring their child’s medical care to ensure optimal health and survival. Many studies suggest that parents of children with CHD are more likely to experience elevated symptoms of depression, anxiety, stress, and anger, as well as poorer QOL.53–57 Research has shown that parental anxiety about raising a child with CHD is more directly related to the diagnosis per se and not the severity of the defect.31,32,53,54,56,58 In addition, the quality of the parent-child relationship, and not disease severity, holds the key to successful transition, with perceived parental acceptance having a more positive effect on adolescent psychological well-being.33,59

Parental anxiety can lead to overprotection and may also explain parents’ reluctance to shift greater responsibility for illness management to their adolescent.27,60 Many adults with CHD recall parental overprotection during their teenage years.32,51,61,62 Parenting styles can either hinder or support the appropriate integration of illness into lifestyle.63 Furthermore, even after parents have relinquished control over their adult child’s healthcare management, control might shift back to the parents in situations when there is dissatisfaction with medical communication or competency of care.62 McDonagh64 highlighted the importance of “negotiating” parental involvement throughout the transition process.

Parent-adolescent perceptions of QOL may not always be in agreement. Casey et al65 found that parents underestimated their child’s physical exercise tolerance in 80% of cases, and in their study on QOL in children with heart disease, Uzark et al16 found that parent-adolescent agreement about QOL was consistently lower for the adolescent group than for their younger counterparts, with the greatest difference occurring in the domain of social functioning. Other studies, however, have found that children and adolescents with CHD reported more behavioral/emotional problems and poorer QOL than their parents reported about them.66,67 To fully understand the impact of CHD on adolescents, therefore, it is important to gather information from both teens and their parents during the transition process. Optimal transition involves collaboration; in addition to patients and healthcare providers, parents and other family members must be integrated into the process.

There are limited data on the impact of CHD on other family members. Janus and Goldberg68 looked at the impact on healthy siblings. High treatment intensity was associated with high accommodation of illness and elevated behavior problems among patients. In contrast, siblings in families in which treatment intensity was low but accommodation of illness was high were at most risk for behavior problems.68 On the basis of data from a parental survey, Wray and Maynard69 reported that a having a child with CHD was perceived to have a negative impact on many areas of family life for approximately one fifth of the sample, particularly in those families in which the child was perceived to be more ill. Family relationships, however, were affected in a very different way, with 43% reporting that family members had become closer, and only 8% reporting that they had been “pulled apart” by the condition of their child.69

Recommendations

For Adolescents

1. The timing of transition should be guided by emotional maturity and developmental level (as opposed to chronological age) for transition planning (Class I; Level of Evidence C).
2. The adolescent should be engaged in transition planning (Class I; Level of Evidence C).
3. The adolescent should be asked about their understanding of their disease in relation to their current health status, restrictions on activities, and future goals (Class I; Level of Evidence C).
4. The adolescent should be encouraged to share concerns about QOL issues (physical restrictions, school, peers, social relationships) (Class I; Level of Evidence C).
5. The adolescent’s fears and concerns should be acknowledged in an empathetic, nonjudgmental manner (Class I; Level of Evidence C).
6. The pediatric cardiology provider should initiate and work together with the adolescent on a transition plan using a transition resource binder and/or health “passport” (Class I; Level of Evidence C).
7. Providers should begin to direct health discussions more toward the adolescent than the parent (Class I; Level of Evidence C).
8. QOL issues should be discussed privately with the adolescent (Class I; Level of Evidence C).
9. Be flexible (Class I; Level of Evidence C).

For Parents

1. The pediatric cardiology provider should initiate discussions on transition planning and partner with parents in the process (Class I; Level of Evidence C).
2. The pediatric cardiology provider should solicit information about parental perceptions of their child’s QOL (Class I; Level of Evidence C).
3. The pediatric cardiology provider should encourage discussion of parental understanding of their child’s disease and concerns in relation to future goals (illness management, education, career) (Class I; Level of Evidence C).
4. The pediatric cardiology provider should be nonjudgmental and empathetic when acknowledging fears70 (Class I; Level of Evidence C).

Health Supervision Issues

Primary Care and Medical Follow-Up Needs

The primary care provider can play an important role in the process of transition. Nearly all individuals born with congenital heart defects require both primary medical care and routine cardiac follow-up care. Many types of providers, including family practice physicians, general pediatricians, adolescent medicine specialists, internists, social workers, nurses, and nurse practitioners, can function in the role of primary care provider. Ideally, the number of transfers among primary care providers should be minimized. The coordination of care between primary and specialty providers is particularly important during adolescence, when new medical and social issues arise that interact with the CHD and can affect health status. Expertise in adolescent medicine is important, including sexuality, contraception and pregnancy, drug and alcohol use and abuse, smoking, mental health, and exercise and physical activity. Although discussions should involve the patient and the parents, time must be allotted to discuss all issues with the adolescent alone. Once a patient reaches adulthood, planned transition of primary care to an internist must occur without gaps in primary care. Some period of overlap of care between pediatric/adolescent specialists and internal medicine specialists is ideal, with consideration of not having the patient transfer to adult primary and cardiac care providers at the same time.

Role of the Primary Care Provider

All individuals with chronic lifelong healthcare needs should have access to a “medical home” as defined by the American Academy of Pediatrics’ medical home policy statement.71 According to this policy statement, the primary physician must provide primary, family-centered care. The primary care physician should also maintain an accessible, comprehensive, central record that contains all pertinent information about the patient, preserving confidentiality. The coordinating physician should transmit appropriate primary care and subspecialty reports to appropriate consultants and to the patient and the patient’s family. It is critical to avoid having multiple specialists functioning independently without knowledge of each other’s activities. Patient and family involvement and education act as a safeguard against fractionation of medical care. Careful attention to an accurate and up-to-date patient “passport” is an important component of this. The primary care provider must coordinate care with the CHD expert on a wide variety of noncardiac issues. This need for coordination of services is especially important for patients with multiple medical problems, which is often the case in adolescents and adults with CHD.

The population of adolescents with CHD is widely heterogeneous. They have residua and sequelae of their disease that at the same time may be lesion specific but also variable in severity and manifestations among individuals with similar diagnoses. To appropriately triage a patient with new symptoms or clinical findings, the primary care physician must know the diagnosis, the surgical history, and the current cardiac status, as well as current medications. The primary care provider should be aware of the patient’s baseline functional status and recommended restrictions and have an understanding of the cardiac prognosis. In addition, the primary care provider should recognize what signs or symptoms are potentially related to the cardiac condition and what might represent worsening of the patient’s condition. The primary care provider should be encouraged to contact the cardiologist for any potentially concerning findings.

Medical Follow-Up

Many of the specific issues of medical follow-up, including noncardiac medical problems and surgical needs, psychosocial issues, anesthetic issues, medications and drug interactions, anticoagulation, antibiotic prophylaxis and dental care, exercise and sports participation, contraception and pregnancy, new symptoms or acute illness, travel, education, employment, insurance, diet, and genetic counseling, are addressed in this scientific statement.

Patients with CHD may have other medical problems and may be taking medications with potential drug interactions. For such patients, the primary care physician should be aware of all medications and should be alert to any potential interactions. Examples include drugs that raise serum levels of other drugs or that alter the activity of another drug (e.g., warfarin). Some drugs have direct cardiovascular effects (e.g., tricyclic antidepressants prolong the QT interval) and may be contraindicated in some cardiac patients.

There are published recommendations about the need for and administration of prophylactic antibiotics to prevent bacterial endocarditis.72 The cardiologist should inform the pediatrician, dentist, and family of the need for such prophylaxis. Nonmedical procedures, such as body piercing and tattoos, that engender a risk of endocarditis73 should be discussed with the patient, family, and pediatrician. In general, such procedures should be discouraged.

Role of the CHD Expert in Medical Follow-Up

To ensure continuity of care, the cardiologist should prepare and maintain an up-to-date, comprehensive cardiac record and transmit that record to the coordinating physician and the patient.74 The record should include stratification of risk and complexity, as well as identification of specific issues that place the patient at increased risk. This information should also be available to the parents and the adolescent patient. The appropriate cardiac information should be updated at the time of each cardiology encounter, and new information should be transmitted to the coordinating physician and the patient.

For successful transition to adult congenital cardiac care, the pediatric cardiologist should prepare a written adolescent transition plan that includes a cardiac destination. The plan should be shared with the coordinating physician and eventually the adult congenital cardiologist. Each pediatric cardiologist should identify the ACHD center to which the transfer of patients will be made.9,21,75,76 In addition to provision for adult cardiac care, the pediatric cardiologist should coordinate transition care with the primary care physician to ensure continuity of primary medical care into the adult healthcare
system. In cardiology encounters during adolescence, the pediatric cardiologist should address medical issues specific to adolescence as they apply to cardiovascular disease. The natural and postoperative history of CHD changes with each decade; thus, even for seemingly simple issues, management evolves with the availability of new outcome data. The cardiologist must be responsible for the ongoing education of the primary care provider and the patient, including education and oversight of the transition process. The cardiologist should also provide guidance to other subspecialists who may have limited experience with CHD to avoid unnecessary testing and misunderstanding.

**Recommendations**

1. The patient’s “medical home” should reside with the primary care provider, who should provide family- and patient-centered care (Class I; Level of Evidence C). The primary care physician should maintain a confidential central record that contains all pertinent information about the patient (Class I; Level of Evidence C).
2. The pediatric cardiologist should prepare a written adolescent transition plan that includes a cardiac destination (Class I; Level of Evidence C).
3. The ACHD expert should prepare and maintain an up-to-date comprehensive cardiac record and transmit that record to the primary care provider (Class I; Level of Evidence C).

**Residual Surgical Issues**

**General Considerations**

Many patients with repaired CHD have residual hemodynamic derangements. The transition process must include education about the likelihood of progression and the potential need for future medical, catheter-based, and surgical intervention. Patients should be alerted to signs and symptoms that suggest worsening status and the need for routine and nonroutine diagnostic assessment. This discussion should focus on the present state of the art and on future developments that may expand available therapeutic options. The patient should be referred to appropriate resources to allow them to remain informed on the latest diagnostic and therapeutic options available to them.

Another consideration is the provision of inpatient care, which may depend on whether the service functions within a stand-alone children’s hospital or a combined medical center. Where to best provide care for the older adolescent or young adult should take into account the resources of the institution, the patient’s medical home, and patient-centered care (Class I; Level of Evidence C). The primary care physician should maintain a confidential central record that contains all pertinent information about the patient (Class I; Level of Evidence C).

**Medical Considerations**

A center that provides services to the ACHD population must have, in addition to experienced surgeons, an active electrophysiology/cardiology team that can provide diagnosis and medical, catheter-based, and intraoperative expertise in the treatment of arrhythmias. The cardiologist and surgeon must recognize the risk of embolic events from thrombus formation within previously placed synthetic systemic-to-pulmonary artery shunts or synthetic patches, recognize the need for bridging of anticoagulation/antiplatelet medications at the time of procedures, and anticipate the presence of acquired heart disease (congestive heart failure, coronary artery disease, valvular disease) over and above the substrate of existing CHD.

The decision to proceed with surgery for a residual or recurrent lesion in an adolescent or young adult with CHD is often based on the assessment of myocardial and valvular function, which is not always straightforward, especially for right and single ventricles. Comprehensive assessment can include echocardiography with newer modalities, cardiac magnetic resonance imaging, and cardiopulmonary exercise testing and arrhythmia monitoring, as well as a detailed history and physical and biomarkers.

**Surgical Considerations**

Special consideration must be given to adolescents and young adults with CHD who present for surgical intervention. Special consideration must be given to adolescents and young adults with CHD who present for surgical intervention. In all cases, catheter-based and surgical options should be discussed thoroughly with the patient. Operative approaches may be altered to address cosmetic concerns without sacrificing optimal visualization of the defects to be repaired. Sequelae of CHD that may impact management and surgical options include hematologic disorders from chronic cyanosis, aortopulmonary collateral development, volume overloading or ventricular failure from intracardiac shunting, and arrhythmias. There are also inherent risks associated with reoperation and resternotomy, that is, chamber, vessel, or conduit entry and phrenic nerve injury. Synthetic pericardial membrane closure during initial surgical interventions may aid in reduction of adhesion formation. Elective procedures should be discussed with the adolescent, and the patient should be given the opportunity to participate in the decision about scheduling, taking into consideration the patient’s academic, athletic, and social schedules. This also enables the patient to participate in medical decision making and develop necessary skills for the future.

An important consideration is the optimal environment for perioperative care, which may require transitioning from an inpatient pediatric service to an adult service. It is important to individualize these decisions depending on the patient’s physiology, the type of operation, and the emotional maturity of the patient.

**Specific Lesions Requiring Intervention**

The majority of adolescent and young adults going through the transition process have already undergone reparative intervention. The number of patients with unrepaired or palliated CHD has diminished as corrective cardiac surgery in infancy has become the standard for many lesions. For example, single-ventricle pathways are often completed before the age of 3 years. Interventional cardiac catheterization techniques have also advanced. Most atrial septal defects, patent ductus arteriosus, and some ventricular septal defects are now closed primarily by use of transcatheter interventions. Initial surgery in the adolescent and young adult population is therefore most likely to be required in those with lesions that are not amenable to percutaneous repair, such as sinus venosus atrial septal defects or partial anomalous pulmonary venous drainage, or in those with progressive...
disease such as aortic root dilatation or progressive stenosis or insufficiency in patients with bicuspid aortic valve. Reoperation is common as patients require new implants (valves, conduits, or pacemakers) or replacement of existing implants because of deterioration or growth of the patient.

Recommendations

1. Follow-up studies, both noninvasive and invasive, should be performed by providers with expertise in the performance and interpretation of the data specific to CHD (Class I; Level of Evidence C).

2. Regular (at least annual) follow-up is required to ensure appropriate hemodynamics are maintained and to prevent secondary complications (Class I; Level of Evidence C). This will reinforce to the patient that regular follow-up is essential indefinitely. Follow-up schedules specific to each disease are discussed in the “ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease.”

3. Nonsurgical and surgical interventions must be tailored to the individual patient and should be performed in centers with specific expertise (Class I; Level of Evidence C).

Electrophysiology/Pacemaker Issues

In adults with repaired CHD, the development of symptomatic arrhythmias is the most frequent cause of hospital admissions and is associated with significant morbidity, including exercise intolerance, heart failure, and thromboembolic complications including stroke and syncope, as well as mortality. Although late ventricular arrhythmias have received the most attention from medical personnel, more recent data show that the development of late atrial arrhythmias is far more common and important because of atrial thrombus formation.80,83

Atrial septal defects, both repaired and un repaired, have a high incidence of developing supraventricular tachycardias over time, especially among those with late repair.80,83 Such patients and others who have undergone atrial surgery, such as Senning- and Mustard-type repairs for transposition of the great arteries, may be treated successfully with catheter ablation procedures84 or pacemakers with antitachycardia pacing capability. Atrial tachycardia is common in patients with single-ventricle repairs and is poorly tolerated. In these patients, ablation is challenging and has a lower success rate. Ventricular arrhythmias, including premature ventricular contractions and nonsustained ventricular tachycardia, occur in as many as 35% of patients with repaired tetralogy of Fallot or double-outlet right ventricle.85 The risk of sustained ventricular tachycardia or late sudden death approximates 6% to 7% with long-term follow-up.86

Advances in device therapy provide a wide spectrum of treatment options for patients with CHD and arrhythmias that require pacing. Abdominal generator implantation may lessen the external appearance of pacemaker implantation in young patients, which poses a psychological issue for adolescents. Activity restrictions are relatively minor but include avoidance of blunt contact sports such as tackle football, judo, karate, and wrestling. Limitations of venous access and the need for lead and generator replacement with age favor an epicardial approach for pacemaker implantation.

Noncardiac Surgery

The presence of CHD places most patients at increased risk for noncardiac surgery (Table 5). In the transitional period, several additional issues arise in perioperative management. Adolescence and young adulthood may be a period when there is an increased need for surgical interventions. Examples include worsening orthopedic conditions, such as progressive scoliosis, and removal of impacted molar teeth. Even relatively minor surgeries such as tooth extraction may place the complex cyanotic patient at risk for complications.

During the transitional period from the care of a pediatric cardiologist to an adult congenital cardiologist, critical issues for intraoperative management may be overlooked. As in the child or the adult, high- or moderate-risk surgeries in adolescent patients with complex CHD, including those with pulmonary vascular disease, should be performed in hospitals with specialized centers for their care whenever possible. The need to transfer the patient to a tertiary facility may be faced by community physicians who do not have the unique resources required for the perioperative care of these patients. In tertiary centers, the pediatric cardiology and adult cardiology inpatient services may be physically separated, and a decision about the optimal location for the procedure will need to be individualized.

Ideally, elective surgery will be preceded by preoperative planning that includes providers with representation from the adult congenital or transitional heart disease, anesthesiology, and surgical services. The purpose of this planning session should be to address the preoperative evaluation and plan the intraoperative and postoperative management, with an emphasis on the risk factors for adverse surgical outcome. The level of risk for noncardiac surgery depends on the patient’s underlying congenital lesion and ventricular function, as well as the presence and severity of cyanosis and pulmonary hypertension. The urgency and complexity of the surgical procedure also impact risk.

The Surgical Procedure

There are no available publications describing the most common noncardiac operations performed in the adolescent,
There are a number of preoperative management issues that need to be addressed in the patient with CHD. Consultation should be obtained from a cardiologist experienced in the care of adolescents with CHD. In general, most cardiac medications should be continued until the time of surgery and resumed as soon after surgery as possible. Patients may require substitution of warfarin with heparin depending on the indication for anticoagulation. In patients with mechanical prosthetic valves or classic Fontan surgery (direct right atrium–to–pulmonary artery connections), the thrombotic risk is high, and the period without anticoagulation should be minimized. In the cyanotic patient with marked degrees of secondary erythrocytosis, a bleeding diathesis has been recognized. Preoperative phlebotomy has been recommended to reduce bleeding risk in some cyanotic patients with secondary erythrocytosis and hematocrit greater than 65%.16,91 Procedures likely to be associated with bacteremia require endocarditis prophylaxis according to the most recent AHA guidelines, “Prevention of Infective Endocarditis.”92

Intraoperative Management Issues
The anesthesiologist must pay careful attention to the physiological consequences of mechanical ventilation and the fluid balance that is required in patients with complex CHD.93,94 For example, in patients with single ventricles and Fontan physiology, the pulmonary blood flow is passive and in patients with Fontan-type surgery), serum albumin levels are low.

Liver function tests (especially in the presence of right-sided heart failure and in patients with Fontan-type surgery), serum albumin levels

Renal function
Arterial blood gases
Pregnancy test (in appropriate females)

CBC indicates complete blood count.
Table 9. Preoperative Cardiac Evaluation

| ECG for evidence of conduction system disease and sinus node dysfunction. |
| Congenitally corrected transposition of the great vessels (l-TGA) |
| Transposition of the great vessels (d-TGA) after atrial switch procedures (Mustard or Senning operations) |
|1-TGA indicates levo-transposition of the great vessels; d-TGA, dextrotransposition of the great vessels.|
| Atrial/ventricular septal defects (endocardial cushion defect) |
| Holter monitor or event monitor in patients with symptoms suggestive of arrhythmias |
| Echocardiogram to evaluate |
| Ventricular size and function |
| Systemic right ventricular function in patients with l-TGA and d-TGA after atrial switch |
| Residual shunts |
| Severity of valvular disease |
| Pulmonary hypertension |
| Cardiac catheterization: Indicated when noninvasive studies are inconclusive with regard to pulmonary arterial pressure, ventricular function, and severity of valvar obstruction |
| Pacemaker/defibrillator interrogation |

and in cyanotic patients with elevated hematocrit. Urine output, central venous pressure, and intraoperative blood loss must be monitored carefully in these patients. In certain cases, a pulmonary artery catheter should be considered for monitoring left-sided filling pressures. Intraoperative transesophageal echocardiography is an alternative method for the evaluation of ventricular filling and function.

Particular care must be exercised to avoid introduction of air into intravenous lines and central catheters to minimize the risk of air embolism, especially in patients with Eisenmenger physiology. The choice of anesthetic agent should be individualized, with consideration of the cardiodepressant effects of some of the inhalation agents and the vasodilating properties of other agents. Intraoperative management of pacemakers and defibrillators requires consultation from the electrophysiologist. If at all possible, use of electrocautery should be avoided. When it is required, the pacemaker-dependent patient should be programmed to the asynchronous mode to avoid interference.95 Other precautions include the use of a bipolar cautery system and assurance that the current path for the cautery does not include the pacemaker. Laparoscopic procedures may be especially hazardous in the patient with CHD. Insufflation of the abdomen may cause a decrease in venous return with a reduction in cardiac output. There is also a potential for paradoxical gas embolism in patients with residual shunts.

Postoperative Management Issues

Fluid management may require postoperative invasive monitoring in the patient with tenuous hemodynamic status. Abdominal surgery may be associated with considerable third-space losses that persist for several days postoperatively. With continued postoperative mechanical ventilation, the intensivist needs to be aware of the potential adverse effects of positive end-expiratory pressure and high tidal volumes. Once the patient is extubated, early mobilization and attention to pulmonary toilet may reduce the risk of postoperative pulmonary infection. In general, most cardiac medications, including anticoagulants, should be started as soon after surgery as possible. Occasionally, in the postoperative patient, equivalent doses of parenteral drugs may be substituted for the oral preparation.

Appropriate pain management is important for patient comfort and to minimize the hemodynamic demands placed by tachycardia and hypertension associated with pain. On the other hand, hypoventilation associated with excessive narcotic use may lower oxygen saturation and increase pulmonary vascular resistance. Adolescents and young adult patients with CHD may have lower thresholds for pain tolerance, influenced by childhood experience. Preoperative evaluation of the patient’s perception of pain tolerance may be helpful in the postoperative period.

Recommendations

1. High- or moderate-risk surgeries in adolescent patients with complex CHD should be performed in hospitals with specialized centers for the care of patients with CHD (Class I; Level of Evidence C). Depending on a number of factors, the choice of setting (pediatric or adult inpatient) should be individualized (Class I; Level of Evidence C).

2. Elective surgery should be preceded by preoperative planning that includes ACHD experts, as well as anesthesia and surgical services (Class I; Level of Evidence C). Original operative and catheterization reports should be obtained and reviewed (Class I; Level of Evidence C).

3. Intraoperative management should be performed by an anesthesiologist familiar with the physiology imposed by the patient’s CHD (Class I; Level of Evidence C).

Follow-Up Needs for Diagnosis/Repair

As noted in the 32nd Bethesda Conference, there are nearly 1 million adults with congenital heart defects in the United States. Despite extremely successful diagnostic and treatment strategies, more than half of these adults have complex conditions or residual issues that require follow-up and treatment by physicians with experience and expertise in the care of these patients. These adults should have their care coordinated with a regional ACHD center, as defined in the guidelines from the 32nd Bethesda Conference, and each patient should have a primary care physician who has 24-hours-a-day, 7-days-a-week backup from a regional center to provide optimal ongoing care. If the ACHD center is not within 1 to 2 hours’ travel, a cardiologist with experience in CHD who is in the patient’s local area and who can help with urgent patient issues should be involved. A detailed approach to each lesion is provided in the ACHD guidelines.16

General principles of coordinating care are depicted in Tables 10 and 11. Table 10 includes health maintenance needs of particular importance to CHD patients. Common medical issues in cyanotic patients of relevance to both the primary care physician and the cardiologist are listed in Table 11. Thorough review of both prescribed and over-the-counter
Table 10. Health Maintenance Needs Usually Handled by Primary Care Provider

- Vaccinations
- Cholesterol screening
- Hypertension screening
- Cancer screening
- Assessment of tobacco, alcohol, and drug use
- Nutritional counseling
- Contraception, sexuality issues

medications is important to assess for drug interactions and potential adverse reactions.

Nearly all patients with repaired defects or mild defects can engage in moderate- to high-intensity exercise. Patients with complex defects, cyanosis, ventricular dysfunction, or symptomatic arrhythmias require evaluation by a specialist in ACHD for an individualized exercise prescription. The report from the 36th Bethesda Conference and ACHD guidelines give detailed descriptions of individual lesions and recommendations for sports participation and can be used as guidelines.

Anticipatory Guidance

Genetic Counseling

Although approximately 1 in 100 infants is born with a congenital heart defect, the genetic cause and recurrence risk vary. As the adolescent approaches the reproductive years, the cause of the congenital heart defect and inheritance patterns should be addressed if these issues have not been addressed previously. These investigations can potentially define the risk to the adolescent’s offspring, and the information can be provided at an appropriate developmental stage. A genetics evaluation, with a thorough clinical evaluation and newer diagnostic tests, may offer diagnostics previously unavailable to families and their care providers.

CHD can occur in isolation (nonsyndromic) or as a component of an underlying genetic or cytogenetic anomaly (syndromic). It may also occur secondary to environmental factors such as diabetes mellitus or prenatal exposure to alcohol, or it may be the result of multifactorial inheritance.

Table 11. Medical Issues in Cyanotic Patients

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Erythrocytosis</td>
</tr>
<tr>
<td>Compensated (stable hematocrit, iron replete, symptoms of hyperviscosity mild or absent)</td>
</tr>
<tr>
<td>Decompensated (rising hematocrit, iron deficient, symptoms of hyperviscosity severe)</td>
</tr>
<tr>
<td>Cholelithiasis</td>
</tr>
<tr>
<td>Abnormal hemostasis</td>
</tr>
<tr>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td>Hemorrhage</td>
</tr>
<tr>
<td>Renal dysfunction</td>
</tr>
<tr>
<td>Hyperuricemia and gout</td>
</tr>
<tr>
<td>Hypertrophic osteoarthropathy</td>
</tr>
<tr>
<td>Scoliosis</td>
</tr>
</tbody>
</table>

Approximately 10% to 15% of newborns with CHD will have an underlying genetic basis for the disease. The majority of cases are considered to be a result of multifactorial inheritance.

For these adolescents, the process of delineating the cause of the heart disease and its impact on their offspring can be instrumental in their understanding of risks for their children. In some cases, the risk to their offspring will be no higher than that for the general population; in other cases, it may be as high as 50%. For some affected individuals, the risk may be not only for recurrence of a congenital heart defect but for a range of birth defects or disabilities associated with an underlying genetic syndrome.

To assist the clinician in evaluating the mode of inheritance for a specific family, a number of items should be taken into consideration. A checklist recommended for the adult patient with CHD can also be implemented in the adolescent years. Recommendations include an evaluation for other medical conditions; assessment of noncardiac malformations and facial dysmorphism; determination of cognitive ability; confirmation of prior genetic studies; and completion of a detailed family history. The information obtained can be used to identify families who can benefit from referral to a genetic specialist. Genetic evaluation of an adolescent with CHD requires a multifaceted approach that often entails the input of multiple specialists. Elements of the process should include the following:

- Targeted 3-generation family history, including individuals with CHD, other malformations, pregnancy loss, or developmental delay
- Genetic evaluation: Syndromic versus nonsyndromic disorder
- Cytogenetic and/or molecular testing if indicated

For some adolescents, further counseling about risk of recurrence and reproductive counseling about prenatal screening and other diagnostic options may need to be delayed until there is appropriate emotional maturity, but these should be offered to the pregnant adolescent. The ACHD specialist should assess the patient’s understanding of the information provided by the genetic counselor and its emotional impact. Counseling should be individualized, with consideration given to the patient’s emotional maturity and family dynamics. In many cases, it may be preferable to include a parent/guardian or other family member of the patient’s choice.

An important resource that has been beneficial to geneticists is available to all practitioners. GeneTests (www.genetests.org) is a service that provides information about laboratories that offer specific testing for specific disorders. Reimbursement for these tests varies by test and insurance carrier. Gene reviews on important syndromes can also be found at this Web site. Another helpful tool is Online Mendelian Inheritance in Man (www.ncbi.nlm.nih.gov/sites/entrez?db=omim), which now lists 363 genetic disorders that include a congenital heart defect as a presenting finding. Many other resources are available that describe the increasing number of cardiac defects associated with single gene disorders/cytogenetic syndromes and multifac-
Sable et al  Best Transition Practices for Adolescents With CHD  1467

Table 12. Empirical Recurrence Risk for Isolated CHD 102

<table>
<thead>
<tr>
<th>CHD</th>
<th>Offspring Affected, %</th>
<th>Father Affected, %</th>
<th>Mother Affected, %</th>
<th>One Sibling Affected, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>4</td>
<td>2–3</td>
<td>6–10</td>
<td>3–6</td>
</tr>
<tr>
<td>ASD</td>
<td>4</td>
<td>1–3</td>
<td>4–11</td>
<td>2–3</td>
</tr>
<tr>
<td>TOF</td>
<td>4</td>
<td>1–5</td>
<td>2–4</td>
<td>2–3</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>6</td>
<td>2</td>
<td>4–6.5</td>
<td>2</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>3</td>
<td>2–2.5</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>5–10</td>
<td>3–5</td>
<td>13–18</td>
<td>2–3</td>
</tr>
</tbody>
</table>

CHD indicates congenital heart disease; VSD, ventricular septal defect; ASD, atrial septal defect; and TOF, tetralogy of Fallot.

Ethical considerations for the adolescent should not be ignored, because potential stigmatization can be a concern when discussing genetic disorders. In some cases, additional support may be beneficial, including social workers and psychologists. It may be difficult for adolescents to understand and incorporate how information about their genetic makeup can affect their future family. The approach to these topics should take place in an environment where psychological counseling or other support is available, if needed.

Recommendations

1. All patients should have a targeted 3-generation family history that includes individuals with CHD, other malformations, pregnancy loss, and developmental delay (Class I; Level of Evidence C).
2. Genetic evaluation with cytogenetic and/or molecular testing should be performed if indicated (Class I; Level of Evidence C).
3. Recurrence risk and reproductive counseling, including prenatal screening and/or diagnostic operations, should be presented to adolescent patients who are pregnant (Class I; Level of Evidence C).
4. Counseling should be individualized, with consideration of the adolescent patient’s emotional maturity and potential impact of counseling (Class I; Level of Evidence C).

Sexuality, Pregnancy, and Reproductive Issues

Introduction

Although the primary responsibility for discussion of sexuality and reproductive issues should fall to the primary care physician, the cardiologist may have more frequent visits with certain patients, especially those with complex lesions, who require frequent monitoring. Therefore, the cardiologist’s awareness of these important issues is critical to patients’ well-being. Although some studies have shown that adolescents with CHD are less likely to be engaged in sexual behavior, 48, 49 they may be taking greater risks because of a lower level of knowledge about safe practices, with a lower prevalence of contraceptive use than among their counterparts. 103 It may be that their parents are more reluctant to discuss issues of sexuality and reproduction, 104 and as a result, these patients may enter adulthood with many misconceptions and fears about their sexuality and their ability to conceive and bear children. 104, 105 Therefore, it is important for transitional care programs to initiate routine sexual health risk screening and preventive counseling that includes topics such as sexuality, contraception, and reproduction for adolescents with CHD.

Adolescent Sexual Activity

The majority of teenagers report having had sexual encounters by mid to late adolescence. 106 The National Survey of Family Growth provided data that indicated ≈47% of female teenagers and ≈46% of male teenagers had engaged in sexual intercourse at least once. 107 Approximately one fourth of all youth reported having had intercourse by 15 years of age, 107 with the average age for first sexual intercourse occurring being 17 years for girls and 16 years for boys. 108, 109 One study reported higher rates of early sexual encounters among youth with chronic conditions than among those in a comparison group. 110 Emotional distress and depression, which are more prevalent in adolescents with chronic disease, constitute a recognized risk factor for engaging in high-risk behaviors 108, 111; however, these studies were not specific to patients with CHD. Although the importance of addressing sexual practices, contraception, and pregnancy has long been recognized, little is known about the sexual and contraceptive practices of adolescents with CHD. 4 In 1 study of older teens 16 to 20 years of age, however, the overall rates of sexual activity were lower than in a comparison sample. These lower rates were attributed to delays in psychosocial development reported in these patients. 112 Among sexually active patients, 72% of the 16-to-18-year-olds were likely to have engaged in sexual intercourse within the past 3 months, using questionable birth control, or using drugs or alcohol before sex. 49

Contraception

Despite a trend for increased use of some form of birth control among teens at the time of first intercourse, 50% of
adolescent pregnancies still occurred within the first 6 months of the first sexual encounter. The most common method of contraception among female teens is the “pill.” Today, oral contraceptives contain a combination of a low dose of estrogen (20 to 35 μg) with varying amounts of progestin. The low doses of estrogen in the various forms of contraceptives have less effect on platelet activation and clotting than the higher doses of estrogen used previously. However, because of the risk of thromboembolic complications, estrogen-based oral contraceptives are considered contraindicated in females who are cyanotic, those with right-to-left shunts, and those with pulmonary vascular disease. Oral contraceptives are relatively contraindicated in patients who have prosthetic valves or conduits.

Progestin-only contraceptives inhibit ovulation and have no measurable impact on clot formation or platelet function, thus offering an acceptable alternative to estrogen-based contraception. Some of the progestin-related side effects include breakthrough vaginal bleeding, amenorrhea, and a diuretic response. The only oral form available in the United States is the “minipill,” but because of its short half-life, it must be taken at the same time each day, which makes it a poor choice for many adolescents. A backup form of contraception must be used for 48 hours if a pill is missed. Depot medroxyprogesterone acetate (DMPA), known as Depo-Provera, is a long-acting progestosterone-only injectable that is administered every 12 weeks; it is a preferred method of artificial birth control in adolescents for whom compliance is a concern. Unfortunately, the associated side effects (breakthrough bleeding, weight gain, and headache), most of which occur in the first year, contribute to a high discontinuation rate (60%) among teens. Additionally, the risk of osteoporosis associated with amenorrhea with long-term use is of concern. Although there are no specific guidelines for teens, it is generally recommended that females who use this method for >2 years or who smoke or are extremely thin should be monitored regularly for bone density loss and be prescribed calcium supplements (1200 to 1500 mg/d). The contraceptive implant Implanon is a favorable choice for teens with compliance issues or for patients for whom pregnancy is contraindicated but who are not yet ready to undergo permanent sterilization. Implanon consists of a single small plastic rod that is effective for 3 years and has lower risk with insertion.

Although an effective contraceptive method, the intrauterine device may not be considered an appropriate first choice for adolescents, primarily because of the risk of sexually transmitted disease. With proper counseling, however, this may be a viable method in selected patients. Barrier methods have the highest failure rate because of inconsistency of use and the discomfort young people may feel when applying the device. The diaphragm is the most commonly recommended barrier method of female contraception in adolescents. Used with spermicide, it is reported to be 80% effective. Regardless of the contraceptive method selected, the addition of a male condom with spermicide must be stressed to ensure protection against sexually transmitted diseases, HIV, and hepatitis B.

Emergency contraception is used to reduce the risk of pregnancy after unprotected vaginal intercourse. The 2 most commonly used methods are the combined estrogen-progestin (Preven) pill, referred to as the Yuzpe regimen, and the progestin-only regimens of oral contraceptive pills, referred to as Plan B. Both consist of 2 doses of contraceptive steroids taken 12 hours apart. They can reduce the risk of pregnancy up to 120 hours after unprotected vaginal intercourse but are more effective when the first dose is taken within 72 hours. Plan B, which is available from drugstores without a prescription for females who are ≥17 years of age, is the preferred method for patients at risk for thromboembolic complications.

Because of potential emotional consequences, permanent sterilization is generally not recommended for adolescents. For adolescent females, it is preferable to postpone tubal ligation until they are sufficiently mature to participate in the decision. When there is a decision to proceed with permanent sterilization, a transcervical procedure that uses coils that lead to fallopian tube occlusion is now available. Clinical trials of this device report the fallopian tubes are closed within 3 to 6 months, and thus, additional contraception is required early after the procedure.

Regardless of an adolescent’s previous sexual history, abstinence can always be presented as an option. Because peer pressure plays a key role in the onset of sexual activity, adolescents need to be encouraged to avoid engaging in sex to please others.

Selecting a Contraceptive Method

The selection of a contraceptive method for the adolescent female with CHD must be individualized by taking into account the primary cardiac defect, any related surgical intervention and medication, postoperative residua and sequelae, and the adolescent’s maturity. The World Health Organization has recommended that females who have undergone successful surgical repair for simple lesions such as atrial septal defects, patent ductus arteriosus, and ventricular septal defects with no sequelae have no restrictions in their choice of contraception. Patients with lesions for which estrogen-based contraceptives are contraindicated are outlined in Table 13.

Pregnancy

Adolescent pregnancy is considered high risk from psychological, medical, and sociological perspectives. Psycho logically, pregnancy interrupts normal developmental tasks by forcing teens to assume adult responsibilities before they are prepared to do so. Adolescents are at higher risk for complications of pregnancy because they frequently fail to seek early prenatal care and are often noncompliant with medical recommendations, rendering them at risk for anemia, pregnancy-induced hypertension, preterm births, and low-birth weight infants. For the adolescent with CHD, there is the additional maternal and fetal risk posed by her cardiac condition. The adolescent should be counseled about the teratogenic effect of medications. Social issues such as access to care, health coverage, and ambivalence about informing their parents create barriers to early prenatal health care. Transitional care programs for adolescents should emphasize the importance...
of early counseling and medical evaluation should an unplanned pregnancy occur despite guidance and education on contraception.

For the pregnant adolescent, the laws about parental notification and parental consent for termination vary by state. In several states, a pregnant adolescent is considered an emancipated adult, requiring no parental consent to terminate the pregnancy. Knowledge of the state laws governing a minor’s rights is an important component of the transitional program.

Medical termination is not without its own risks, particularly for the patient with complex CHD. It is therefore important to act swiftly, because the choice of termination procedure is determined by the duration of the pregnancy.128 The emotional impact of pregnancy termination should not be underestimated, and appropriate personnel should be available for counseling, which must be offered in a supportive, nonthreatening environment. For the adolescent who chooses to continue the pregnancy, a well-coordinated pregnancy and delivery plan must be developed, one that includes not only the obstetrician and cardiologist but clinical social work and psychological services personnel to assist and support the patient in making the decision about keeping the baby or placing the baby into adoptive services.

Counseling the Adolescent: Role of the Healthcare Provider

In addition to the topics of contraception and pregnancy, discussions with adolescents should stress prevention of sexually transmitted diseases and HIV. To facilitate these discussions, an environment must be established that is supportive and nonjudgmental and that establishes a level of trust and confidence. Additionally, it is important to encourage parents to discuss sexuality and contraception in a manner consistent with the family’s attitudes, values, and beliefs. Timing of these discussions will vary. Health education materials should be available to preadolescents and their parents. With the onset of puberty, a comprehensive history should include an assessment of the patient’s knowledge about sexuality, sexual feelings, and other high-risk behaviors, including use of alcohol and nonprescription drugs. Additionally, handouts to reinforce safe sex practices and responsible decision making should be available in the office or clinic. Once menarche begins, a gynecologic history should be obtained routinely during cardiac follow-up visits. For adolescents who are contemplating becoming sexually active or who demonstrate high-risk behaviors, referral to a contraception clinic or to gynecologic services should be provided.

Recommendations

1. Introduce age-appropriate reproductive health information on sexual health, contraception, and pregnancy beginning early in adolescence (age 12 years) (Class I; Level of Evidence C).

2. Healthcare practitioners who are knowledgeable in both adolescent sexual and reproductive health and CHD should provide clinic-based teaching and counseling on contraception and pregnancy (Class I; Level of Evidence C).

Table 13. Recommendations for the Use of Combined Hormonal Contraceptives in Women With Cardiac Disease

<table>
<thead>
<tr>
<th>WHO 1 Always Useable</th>
<th>WHO 2 Broadly Useable</th>
<th>WHO 3 Caution in Use</th>
<th>WHO 4 Do Not Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Condition with no restriction for the use of the contraceptive method</td>
<td>Condition for which the advantages of the method generally outweigh the theoretical or proven risks</td>
<td>Condition for which the theoretical or proven risks usually outweigh the advantages of using the method</td>
<td>Condition represents an unacceptable health risk if the contraceptive method is used</td>
</tr>
</tbody>
</table>

- MVP with trivial MR
- Bicuspid aortic valve with normal function
- Mild PS
- Repaired coarctation with no hypertension or aneurysm
- Simple congenital lesions successfully repaired in childhood and with no sequelae (atrial or ventricular septal defect, patent ductus arteriosus, or total anomalous pulmonary venous drainage)
- Most arrhythmias other than atrial fibrillation or flutter
- Uncomplicated mild native mitral and aortic valve disease
- Tissue prosthesis valve lacking any of the features noted in WHO 3 or 4 columns
- Surgically corrected CHD lacking any of the features noted in WHO 3 or 4 columns
- Small left-to-right shunts not reversible with physiological maneuvers (ie, small ventricular septal defect, small patent ductus arteriosus)
- HCM lacking any WHO III or IV features
- Past cardiomyopathy, fully recovered, including peripartum cardiomyopathy
- Uncomplicated Marfan syndrome
- Small left-to-right shunt not reversible with physiological maneuvers
- Atrial fibrillation or flutter on warfarin
- Bileaflet mechanical valves in the mitral or aortic position taking warfarin
- Atrial septal defect with left-to-right shunt that may reverse with physiological stress (eg, Valsalva maneuver)
- Repaired coarctation with aneurysm and/or hypertension
- Marfan syndrome with aortic dilation unoperated
- Previous thromboembolism
- Atrial fibrillation or flutter, if not anticoagulated
- Bjork-Shiley or Starr-Edwards valves even taking warfarin
- Dilated left atrium >4 cm
- Fontan heart even taking warfarin
- Cyanotic heart disease even taking warfarin
- Pulmonary arteriovenous malformation
- Past thromboembolic event (venous or arterial) not taking warfarin
- Prior LV dysfunction due to any cause (eg, dilated cardiomyopathy), LVEF <30%
- Coronary artery disease
- Coronary arteritis (eg, Kawasaki disease with coronary involvement)

WHO indicates World Health Organization; MVP, mitral valve prolapse; MR, mitral regurgitation; CHD, congenital heart disease; PS, pulmonary stenosis; HCM, hypertrophic cardiomyopathy; LV, left ventricular; and LVEF, left ventricular ejection fraction.

3. Young teens should be assessed for evidence of high-risk behaviors that could expose them to sexually transmitted diseases, HIV, and pregnancy, and this information should be included in the patient’s history (Class I; Level of Evidence C).
4. Providers should document prescribed interventions to modify high-risk behavior (Class I; Level of Evidence C).
5. Providers should document prescribed contraception for females and recommended use of condoms for males (Class I; Level of Evidence C).
6. Comprehensive adolescent reproductive services that include adolescent gynecology should be provided (Class I; Level of Evidence C).
7. Educational materials and local resources on sexuality and contraception should be provided for adolescents and their parents, and for females with CHD, the importance of planning all pregnancies should be discussed (Class I; Level of Evidence C).

Exercise Prescription/Rehabilitation
Because more CHD patients survive to adulthood, it is also important to focus on QOL with aging and the onset of adult comorbid conditions. Exercise is an important component of QOL and health maintenance. Although studies of functional capacity have not been undertaken on a large, multicenter scale, a number of smaller studies have been performed. The ages of patients and methodologies used in these studies vary, but most report benefits of exercise and that at least some level of exercise is safe. In general, they also suggest that exercise tolerance is not likely to be “normal” for many patients but that many are also capable of performing some level of regular exercise. Many children are not encouraged to exercise because of parental, school, or personal fears about their underlying heart condition, and thus, the patients do not exercise much as they enter adulthood. This is a cause for concern for overall health as they enter adulthood, because a sedentary lifestyle can contribute to other comorbid conditions such as obesity, coronary artery disease, hypertension, diabetes mellitus, dyslipidemia, and osteoporosis.

Exercise Assessment and Benefits
Exercise is beneficial in overall health for prevention of adult-onset diseases, as well as in recovery from chronic conditions, including coronary artery disease and congestive heart failure. Despite the potential benefits of exercise, it is often not discussed as part of a routine clinic visit. In a study of 99 adults with CHD, exercise was discussed by the physician in only 28 cases, and discussions generally focused on exercises to avoid rather than encouraging appropriate activity. On the other hand, 44% of patients thought all exercise was safe. The most common barriers to exercise described by patients included symptoms and fears. In general, exercise in the CHD patient population has been evaluated by either subjective patient interpretation or objective exercise testing. Subjective studies usually include questionnaires about QOL or interest in exercise participation. The objective measures include those assessed by routine exercise treadmill testing, cardiopulmonary exercise testing including ventilatory parameters, or a 6-minute walk test. These tests provide measurable data such as standardized exercise time, maximal oxygen consumption, or distance walked, respectively. Adverse outcomes, such as electrocardiographic evidence of ischemia or strain, arrhythmias, oxygen desaturation, and hypotension, can be detected. Although important for research, this information may also assist in clinical decision making and risk assessment for sports participation or may be used as a basis for a recreational or rehabilitation exercise prescription. The results can also be reassuring for patients or families who may be afraid to exercise because of their CHD.

Regular exercise among adolescents and young adults with CHD has been evaluated in small, single-center analyses. Small studies of pediatric and adult patients with CHD have shown that exercise or exercise training programs may improve functional capacity and some measures of QOL. In a study by Gratz and colleagues, adolescent and adult CHD patients (14 to 73 years old) reported decreased QOL scores in physical function and general health domains compared with peers, but they did not show decreased scores in psychosocial aspects. Interestingly, they also significantly overestimated actual exercise capacity based on exercise test results. So, although these CHD patients may have limitations, it did not appear to affect their psychosocial function or estimation of their own exercise capacity. In another study, structural and pathophysiological diagnoses were related to exercise ratings, but they were poorly predictive of exercise capacity; thus, other factors likely play a role.

Short-term exercise training programs have been evaluated. Although exercise training or rehabilitation programs for children and adults with CHD are thought to be safe and likely to be effective at improving exercise parameters, the results of small studies of exercise training in children and adults with CHD have shown mixed results. One study using home exercise and dyspnea thresholds in children did not show any improvement. Another study evaluated a 12-week cardiac rehabilitation program in a small number of teenagers with complex heart disease. Even in this short time, improvements in peak oxygen consumption and heart rate recovery were noted. Fredriksen and colleagues showed an increase in peak oxygen consumption and improvement in psychosocial measures after exercise training, including decreased social withdrawal and decreased somatic complaints. Finally, another study that evaluated a 3-month exercise training program in adult patients with repaired tetralogy of Fallot also found an increase in peak oxygen consumption.

Developing good exercise habits in childhood can provide lifelong benefits. The health benefits are clear in terms of decreased risk of obesity and adult-onset cardiovascular disease. There also may be benefits in prevention of other conditions, such as osteoporosis, with the use of mild weight-bearing exercise. It is known that obesity is already a problem in children and young adults in the United States but also specifically in CHD patients. Adolescent lifestyle habits, including exercise, related to atherosclerosis risk were assessed in children with CHD in Belgium. They found that only 15% spent 30 minutes per day in physical activity. Although adolescents with CHD may not practice perfect health-promotion behaviors, one study from Taiwan showed
that they are no different from their peers in that country. The consequences, however, may be greater for the CHD patients, which makes lifestyle choices even more important.\textsuperscript{142} Physical activity was also assessed in adults with CHD in the United Kingdom. Only those in New York Heart Association class I performed similar activity to a sedentary reference population, whereas groups in New York Heart Association class II or III exercised less than half as much as those in New York Heart Association class I. Despite this, patients reported a desire to participate in exercise.\textsuperscript{143}

In any exercise endeavor, it is important that the adolescent CHD patient receive support to succeed and develop good lifelong habits. A more sedentary lifestyle may contribute to decreased exercise tolerance, and this lifestyle in children may be a result of overprotective parents and teachers.\textsuperscript{144} In another study that evaluated perceptions about exercise in a small group of children with CHD, sports and exercise were thought to be beneficial but not valued. The low self-efficacy in this study appeared to be influenced by fear and exclusion.\textsuperscript{145} Interestingly, a study of teens 12 to 18 years of age showed that belief in self-efficacy was more important than severity of CHD in determining exercise or sports participation. The self-efficacy and attitude of the parent toward exercise participation was significantly influenced by the recommendation of the cardiologist.\textsuperscript{146} To encourage physical activity habits, it appears to be important for children and teens to receive appropriate direction and encouragement from the family and cardiologist, as well as coaches, teammates, and friends.

**Type of Exercise**

The majority of available information and opinion leans toward the benefits of exercise for children and adolescent CHD patients; however, determining the appropriate types and levels of exercise remains very important. When determining appropriate and safe types of exercise, many recreational activities, such as walking, swimming, or biking to various levels of exertion, may be appropriate for many patients. Others may want to participate in higher-level or more competitive sports, including at the high school and college level. For this determination, the use of exercise testing can be beneficial to help gauge safety and level of exercise individually. To improve this decision-making process, the AHA has published 2 sets of guidelines about sports participation and involving patients with congenital or genetic heart disease. The 36th Bethesda Conference recommendations outline different types of CHD and repairs and discuss different types of competitive athletics that may or may not be appropriate.\textsuperscript{96} This document also outlines the differences between static and dynamic types of sports to help determine what may be more appropriate for patients with different types of CHD (Table 14).\textsuperscript{147} The AHA developed a second statement on exercise participation for patients with genetic cardiovascular diseases, including Marfan syndrome and hypertrophic cardiomyopathy. These reports can be helpful for both physicians and patients to guide activities to achieve exercise in a safe and comfortable fashion.\textsuperscript{96}

Although most CHD patients can perform some form of exercise or sports and should be encouraged to do so, there are those who will require even further consideration. All CHD patients should be considered individually when it comes to discussing exercise and exercise programs because of the variability even within types of CHD. However, those with heart rhythm devices, dilated aortas, and coronary anomalies deserve special attention. Those with pacemakers

| Table 14. Classification of Sports\textsuperscript{147} |
|---------|---------|---------|---------|---------|
|         | A. Low  | B. Moderate | C. High |
| Increasing Static Component |     |     |     |
| I. Low | (<40% Max O\textsubscript{2}) | Baseball/Softball\textsuperscript{†}, Fencing, Table tennis, Volleyball | Badminton, Cross-country skiing (classic technique), Field hockey\textsuperscript{†}, Orienteering, Race walking, Racquetball/Squash, Running (long distance), Soccer\textsuperscript{†}, Tennis |
| II. Moderate | (40-70% Max O\textsubscript{2}) | American football\textsuperscript{†}, Field events (jumping), Figure skating\textsuperscript{†}, Rodeoing\textsuperscript{†}, Rugby\textsuperscript{†}, Running (sprint), Surfing\textsuperscript{†}, Synchronized swimming\textsuperscript{†} | Basketball\textsuperscript{†}, Ice hockey\textsuperscript{†}, Cross-country skiing (skating technique), Lacrosse\textsuperscript{†}, Running (middle distance), Swimming, Team handball |
| III. High | (>70% Max O\textsubscript{2}) | Body building\textsuperscript{†}, Downhill skiing\textsuperscript{†}, Skateboarding\textsuperscript{†}, Snowboarding\textsuperscript{†}, Wrestling\textsuperscript{†} | Boxing\textsuperscript{†}, Canoeing/Kayaking, Cycling\textsuperscript{†}, Decathlon, Rowing, Speed-skating\textsuperscript{†}, Triathlon\textsuperscript{†} |

MVC indicates maximum ventilatory capacity; Max O\textsubscript{2}, maximum oxygen consumption.

*Danger of bodily collision.
†Increased risk if syncope occurs. Reprinted with permission from American College of Cardiology Foundation.\textsuperscript{147}
or implantable cardioverter defibrillators may have restrictions as to their maximal heart rates. They also may need to avoid contact sports or have special equipment to protect the device during activity. Those with dilated aortas or connective tissue disease of the aorta also may need to avoid contact sports or high static activity.

**Recommendations**

1. Given the general benefits of exercise for overall health and prevention of obesity, at least some form of routine exercise is advisable for most CHD patients (Class I; Level of Evidence C). Early activity habits will also likely enlist the understanding, support, and encouragement of parents, teachers, and coaches at school.

2. Objective exercise testing should be performed as a baseline in all CHD patients to provide an exercise prescription (Class I; Level of Evidence C). Periodic repeat testing is advised in the presence of new symptoms or other changes in the clinical status or to assess the hemodynamic or arrhythmic response to medication (Class I; Level of Evidence C).

3. On the basis of the results of objective testing, the cardiologist should discuss exercise and physical activity with the patient and family as early in childhood as possible to encourage good habits (Class I; Level of Evidence C). The level of recommended exercise should be reassessed during follow-up and revised according to the patient’s clinical status and the results of repeat testing (Class I; Level of Evidence C).

4. The exercise prescription should include the type of exercise (dynamic or static), heart rate goals and limits (maximum rate if applicable), duration and frequency of exercise, and a guide for advancing the level of exercise (Class I; Level of Evidence C). The prescription should also include exercise to avoid. The exercise prescription should follow previously published guidelines, recognizing, however, that most of these guidelines pertain to competitive sports at a high school and college level (Class I; Level of Evidence C).

5. For patients with limited exercise ability, adverse hemodynamic response to exercise, or recent surgery, a medically supervised cardiac rehabilitation program may be beneficial (Class IIb; Level of Evidence C).

**Education and Career Choices**

An important developmental task for adolescents with CHD is to assume more responsibility for their own health care; however, studies have shown that adolescents and young adults have a poor understanding of their cardiac disease, treatment, and prognosis. Veldman et al reported that only 22% of 7-18-year-olds (mean age 13 years) with CHD interviewed could correctly identify their diagnosis by name, and this was unrelated to age, sex, or severity of the defect. In addition, only 30% had a good conceptual understanding of their disease, with 36% having a wrong or poor understanding of their illness. Moons and colleagues found that although 80% of patients 18 to 46 years of age (mean age 23 years) with CHD were able to correctly identify treatment plans, frequency of follow-up, proper dental practices, appropriate vocational choices, pregnancy risks, and the appropriate use of the contraceptive pill (women), only 50% to 80% were able to correctly identify the name and anatomy of their cardiac lesion; the risks, signs, and symptoms of endocarditis; the genetic risks of CHD; the impact of smoking and alcohol on their heart disease; and the appropriateness of the intrauterine device as a method of contraception (women). Similarly, Dore and colleagues reported that of 104 patients with CHD (16 to 72 years of age) enrolled in their study, the clinical diagnosis was unknown by 34.6% of patients, many of whom had complex CHD. More importantly, although 79% knew about the need for antibiotic prophylaxis, 50% did not know why it was needed.

Rönnings et al used semistructured open-ended interviews to ask 16 adults (19 to 55 years of age, mean age 28 years) about their experiences in learning about their heart disease. The majority of subjects wished that they had received developmentally appropriate education sessions beginning in early childhood, with added information and continuing reinforcement through adolescence. Although few could specifically describe what information should be provided, the subjects did identify the lack of knowledge about their heart disease as a barrier to taking a more active role in decisions about their treatment and care.

Thus, it is common for adolescents and young adults to have significant gaps in their knowledge about CHD, and these deficits must be remedied before we can expect adolescents to become more independent regarding their health care and more responsible for their long-term follow-up.

**Neurodevelopmental Issues**

Academic achievement, learning disabilities, behavioral problems, and inattention/hyperactivity and other psychological issues must be taken into account when planning for transition, because they can all significantly impact an adolescent’s ability to learn and assume responsibility for their health care.

Improvements in the longevity and survival rates for patients with CHD have been accompanied by an increasing awareness of adverse neurodevelopmental outcomes for patients who required cardiac surgery as neonates or young infants. Children with complex CHD such as d-transposition of the great vessels, total anomalous pulmonary venous return, hypoplastic left heart syndrome, and other functional single-ventricle lesions have a significantly higher incidence of problems with academic performance, behavioral abnormalities, inattention, hyperactivity, and the ability to perform higher-order executive functions such as visual-motor integration.

Shillingford and colleagues, in a study on inattention, hyperactivity, and school performance, found that the number of subjects in their study with high risk scores for inattention and hyperactivity was 3 to 4 times higher than that in the general population and that 49% of the 109 enrolled subjects were receiving some type of remedial academic services. Although early research focused on modifying intraoperative technical and procedural factors to prevent and reduce neurological injury, such efforts have resulted in only moderate success. More recent research has pointed to patient-specific...
factors such as low birth weight, age at surgery, length of stay in the intensive care unit, and socioeconomic status as stronger determinants of adverse neurodevelopmental outcomes.154,155

Education and Employment

Education and employment are crucial to financial security and psychological well-being and therefore represent another important concern for these adolescents. Although educational milestones are similar to those of healthy peers, research has shown a significant difference with regard to employment rate. Simko et al156 found full-time or part-time employment in 71% of adults with CHD compared with 84% of healthy subjects (P=0.011). The rate of employment tends to be higher among those with milder and acyanotic forms of CHD.156–158

This high rate of unemployment for adults with CHD is concerning. One possible explanation for this high unemployment rate is the misunderstanding and lack of knowledge about CHD by employers. In a small survey of employers in the United Kingdom, most (10 of 14, or 71%) said they would employ these patients on the basis of current functional assessment, with or without medical opinion, but 4 (29%) of 14 would exclude patients with CHD or would look at the individual’s future health prospects before hiring them. Unfortunately, no consensus about hiring practices exists among employers who hire these young adults, and no actuarial survival statistics are available to help them with their decision-making process.159 When job participation was evaluated, the patients with complex CHD had significantly more job handicaps (primarily mobility related) than patients with mild CHD. Those with complex CHD also tended to cease employment because of physical problems/demands, emotional problems, or problems with transportation, and the most frequent requests for adaptations to the work environment focused on more flexible work hours, reduced time pressure, and increased freedom to organize one’s work.157 These findings underscore the importance of career counseling in these patients to help them find the best fit for their physical and emotional capabilities.

Career Counseling

Numerous challenges await adolescents with CHD as they prepare to enter the workforce. These include discrimination in the workplace, difficulty in maintaining employment during medical crises, and challenges in maintaining or changing employment because of changes in functional capacity.157 Career and education plans should be discussed in early adolescence (13 to 15 years of age) to allow for assessment of the patient’s mental, physical, and social abilities. This assessment and education should involve not only the healthcare provider and patient but also parents, educators, and career advisors. The receipt of structured career/employment advice has been shown to be associated with a higher rate of employment (73%) than no career/employment counseling (46%).158

Discrimination is prohibited in all aspects of the work process, including hiring, advancement, compensation, and job training. It is unlawful for the employer to question an applicant about a disability or require a medical examination unless the information is necessary for specific job-related performance. Employers are also required, barring creation of a substantial hardship, to accommodate and modify equipment and training policies to allow a disabled worker to perform his or her job.160 The US Department of Education’s “Pocket Guide to Federal Help for Individuals With Disabilities is a good resource for information on benefits and services.”161

There are 3 governmental legislative acts that protect patients with disabilities with regard to seeking employment:

- The Rehabilitation Act of 1973 and the subsequent amendment in 1978 prohibit employment discrimination based on disability by any federal employer programs that receive federal funding, federal executive agencies, and the US Postal Service. It also requires that these agencies develop plans for hiring, placement, and education/advancement through vocational training.162
- The second-injury section in worker’s compensation allows an employee to register a preexisting disability with the local worker’s compensation facility. This allows the disabilities or progression of disability to be reimbursed from a special second-injury fund to ensure that employers are protected from future losses.
- The Americans With Disabilities Act is the most comprehensive legislation for individuals with disabilities. It defines disability and extends the scope of protection for these individuals to the private sector. It prohibits disability-based discrimination by any employer who has ≥15 employees for each working day in each of ≥20 calendar weeks in the current or preceding calendar year.163

Parents and patients should be educated about any possible restrictions that might affect ability to work that may develop with age. This education could allow the adolescent to select a career in which full employment can be maintained throughout a lifetime. Motivation does not appear to be a problem with patients with CHD, because more of them proceed to higher levels of education than control subjects.158 Higher levels of education lead to higher levels of job participation.157

In 1986, the AHA published “Recreational and Occupational Recommendations for Young Patients With Heart Disease” that can be used to objectively define and assist with occupational counseling for patients with CHD (Tables 15 and 16).164 Occupational activities should also be assessed for any additional patient-specific risks. For example, isometric exercise increases systemic blood pressure, and activities involving this type of work should be discouraged in patients with left-sided heart lesions such as coarctation of the aorta, aortic stenosis, bicuspid aortic valve, and aortic regurgitation. Patients with left ventricular outflow tract obstructive lesions and ventricular arrhythmias have an increased risk of dizziness and syncope, which may pose grave risks for certain jobs, such as piloting a plane or boat, driving a bus or other public vehicle, or working on high-rise construction projects. A list of occupations and their physical demands is provided by the US Department of Labor.165 Objective measures of

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exercise tolerance should be used to assist in counseling whenever possible.

Once the occupational activity level, education capacity, and social ability have been assessed, the highest level of education should be encouraged. For patients who are handicapped physically or mentally, vocational rehabilitation should be encouraged. Many states offer vocational educational and rehabilitation services. These vocational services perform a full assessment of the individual and can suggest types of employment the individual can be expected to perform. The counselor can also suggest programs that provide appropriate training, and if need be, the patient can apply for financial assistance (“Pocket Guide to Federal Help for Individuals With Disabilities”).

### Recommendations

1. Develop a structured educational/teaching plan based on the adolescent’s individual academic abilities, educational level, and developmental maturity (Class I; Level of Evidence C).

2. Begin counseling early in adolescence to identify interests, as well as the need for additional educational or vocational training and guidance (Class I; Level of Evidence C). Utilize exercise testing as appropriate to assist in determining physical abilities (Class I; Level of Evidence C).

3. View every patient as employable, and tailor career/employment counseling to the adolescent’s physical capacity, as well as mental and psychological disposition (Class I; Level of Evidence C).

### Insurance (Health and Life)

A significant proportion of patients with CHD in the United States experience problems obtaining health insurance or continuing health insurance coverage after reaching independence in adulthood, which will hopefully be addressed through healthcare reform. For most children with CHD, coverage is obtained through government programs or a parent’s policy. Some patients may continue to have coverage through the parent’s insurance company if they are students and/or through Medicaid, State Child Health Insurance Programs, or state Title V Children with Special Health Care Needs programs until their 21st birthday. Unfortunately, there is a significant change in healthcare coverage at the very time that transfer to the adult healthcare system traditionally occurs. In the general population, the lack of insurance is highest from 19 to 26 years of age. Nearly one third of the population in this age group lacks health insurance. Those with incomes <200% of the federal poverty level are 2.6 times more likely to be uninsured than those with higher incomes. Thirty percent of this age group lives below the federal poverty level. Individuals in their early 20s are less likely to have full-time employment or to have the type of employment that offers insurance coverage. Only 28% of the population from 19 to 26 years of age has access to sponsored insurance from their employer or spouse’s employment. Likewise, only 13% of uninsured adults in this age range are eligible for Medicaid or other public coverage under current eligibility rules. Callahan and Cooper have shown that uninsured young adults with a chronic condition are 8 times more likely to have unmet medical needs and 6 times more likely to have no access to routine care than insured young adults. The likelihood of an unmet need due to cost among those with a disability was 35% in this population. Thus, poverty and presence of a disability are among the factors that negatively influence healthcare coverage of young adults with CHD at the most vulnerable time of transition to the adult healthcare system.

Another explanation for lower insurance coverage relates to attitudinal differences. Young adults may be less risk averse than older adults, and the maturation of planning/executive skills may be incomplete in the early 20s. At particular risk are those patients who are not symptomatic or disabled but have cardiac pathology that may progress to permanent disability if not managed proactively. Some patients have elected to avoid clinic visits, cardiac catheterizations, or operations because of the personal financial consequences.

Late in adolescence, a transition must be made to ensure coverage of cardiac and noncardiac care. Healthcare professionals should address the issue of insurability before patients with CHD leave their parent’s policy or lose their eligibility for children’s services. In fact, evidence points to very early intervention (3 months before birth to kindergarten) having a positive effect on the likelihood of the individual in the urban poor population having insurance as a young adult. This is consistent with previous long-term studies that have shown a positive effect of early education on later employment and health. Thus, preparation for successful transition to adult health care should begin at first contact with preparation of the family, early assessment of educational needs, and the initiation of appropriate interventions. Vocational or career counseling of adolescents should con-
realistic physical abilities and the educational potential of the adolescent with CHD. Education or job training should be encouraged. An important avenue for obtaining insurance is through group policies available through employers or professional associations. Larger employers are more likely to offer full coverage because of the larger size of their risk pools. Although recent healthcare legislation, the Affordable Care Act, will mandate that young adults be covered by their parents’ health insurance until age 26, and the insurance industry is already moving in this direction, a substantial number of young adults whose parents do not have access to employer-based insurance will remain uncovered. Patients considered totally disabled according to Social Security disability guidelines (\(10\%\) of patients with CHD) may apply for healthcare coverage under their state’s Medicaid program and may be eligible for Supplemental Security Income.\(^{176}\) It is particularly important to note that the criteria for Social Security Disability Insurance are markedly different for children and adults with CHD; therefore, these changes must be thoroughly understood by the patient and parents well before the patient reaches the age of 18 years. A comprehensive and up-to-date resource for employment and insurance is available on the Healthy and Ready to Work Web site (http://www.hrtw.org). A list of resources to assist patients in confronting insurability issues is also provided by the Adult Congenital Heart Association (www.achaheart.org) under information and resources.

### Table 16. Occupational Recommendations for Young Patients With CHD

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Occupational Activity*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic insufficiency</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>II</td>
</tr>
<tr>
<td>Moderate</td>
<td>III</td>
</tr>
<tr>
<td>Severe</td>
<td>IV</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>II</td>
</tr>
<tr>
<td>Moderate</td>
<td>III</td>
</tr>
<tr>
<td>Severe</td>
<td>IV</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td></td>
</tr>
<tr>
<td>No PVOD</td>
<td>I</td>
</tr>
<tr>
<td>Mild to moderate PVOD</td>
<td>III</td>
</tr>
<tr>
<td>Moderate PVOD</td>
<td>IV</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td></td>
</tr>
<tr>
<td>Congestive (dilated)</td>
<td>V</td>
</tr>
<tr>
<td>Hypertrophic</td>
<td>IV</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td></td>
</tr>
<tr>
<td>Operated, normal BP</td>
<td>I</td>
</tr>
<tr>
<td>Hypertensive</td>
<td>III</td>
</tr>
<tr>
<td>Hypertension</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>II</td>
</tr>
<tr>
<td>Moderate</td>
<td>III</td>
</tr>
<tr>
<td>Mitral insufficiency</td>
<td></td>
</tr>
<tr>
<td>Mild, no cardiomegaly</td>
<td>II</td>
</tr>
<tr>
<td>Moderate</td>
<td>III</td>
</tr>
<tr>
<td>Severe (+/− atrial fibrillation)</td>
<td>V</td>
</tr>
<tr>
<td>Mitral stenosis</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>III</td>
</tr>
<tr>
<td>Moderate</td>
<td>IV</td>
</tr>
<tr>
<td>Severe (+/− atrial fibrillation)</td>
<td>V</td>
</tr>
<tr>
<td>Mitral valve prolapse</td>
<td></td>
</tr>
<tr>
<td>Mild, no symptoms</td>
<td>I</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td></td>
</tr>
<tr>
<td>No PVOD</td>
<td>I</td>
</tr>
<tr>
<td>Mild to moderate PVOD</td>
<td>III</td>
</tr>
<tr>
<td>Moderate to severe PVOD</td>
<td>IV</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>I</td>
</tr>
<tr>
<td>Moderate</td>
<td>III</td>
</tr>
<tr>
<td>Severe</td>
<td>V</td>
</tr>
<tr>
<td>Pulmonary hypertension (idiopathic)</td>
<td></td>
</tr>
<tr>
<td>PA pressure &lt;0.5 systemic</td>
<td>IV</td>
</tr>
<tr>
<td>PA pressure &gt;0.5 systemic</td>
<td>V</td>
</tr>
<tr>
<td>Tetralogy of Fallot, postoperative</td>
<td></td>
</tr>
<tr>
<td>RV pressure &lt;50 mm Hg</td>
<td>II</td>
</tr>
<tr>
<td>RV pressure &gt;50 mm Hg or cardiomegaly</td>
<td>III</td>
</tr>
</tbody>
</table>

(Continued)

### Table 16. Continued

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Occupational Activity*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td></td>
</tr>
<tr>
<td>No PVOD</td>
<td>I</td>
</tr>
<tr>
<td>Mild to moderate PVOD</td>
<td>III</td>
</tr>
<tr>
<td>Moderate to severe PVOD</td>
<td>IV</td>
</tr>
<tr>
<td>Other major defects</td>
<td></td>
</tr>
<tr>
<td>(unoperated or palliated only)</td>
<td>IV</td>
</tr>
<tr>
<td>Other major defects</td>
<td></td>
</tr>
<tr>
<td>(postoperative intracardiac repair)</td>
<td>III</td>
</tr>
<tr>
<td>Cardiac arrhythmias</td>
<td></td>
</tr>
<tr>
<td>Complete heart block</td>
<td>II</td>
</tr>
<tr>
<td>Pacemaker (artificial)</td>
<td>II</td>
</tr>
<tr>
<td>Premature atrial contractions</td>
<td>I</td>
</tr>
<tr>
<td>Premature ventricular contractions with normal heart</td>
<td>I</td>
</tr>
<tr>
<td>Congenital or acquired heart disease</td>
<td>III</td>
</tr>
<tr>
<td>Supraventricular tachycardia</td>
<td>II</td>
</tr>
<tr>
<td>Ventricular tachycardia with normal heart</td>
<td>II</td>
</tr>
<tr>
<td>Congenital or acquired heart disease</td>
<td>IV</td>
</tr>
</tbody>
</table>

Wolff-Parkinson-White syndrome         I

CHD indicates congenital heart disease; PVOD, pulmonary vascular occlusive disease; BP, blood pressure; +/−, with or without; PA, pulmonary artery; and RV, right ventricular.

*For occupational recommendations, the numbers (I, II, III, IV, and V) correspond to the restrictions given in the categories listed in Table 15.

Unless otherwise noted, all restrictions apply regardless of whether the patient has had surgery.\(^{164}\)

Patients considered totally disabled according to Social Security disability guidelines (<10% of patients with CHD) may apply for healthcare coverage under their state’s Medicaid program and may be eligible for Supplemental Security Income.\(^{176}\) It is particularly important to note that the criteria for Social Security Disability Insurance are markedly different for children and adults with CHD; therefore, these changes must be thoroughly understood by the patient and parents well before the patient reaches the age of 18 years. A comprehensive and up-to-date resource for employment and insurance is available on the Healthy and Ready to Work Web site (http://www.hrtw.org). A list of resources to assist patients in confronting insurability issues is also provided by the Adult Congenital Heart Association (www.achaheart.org) under information and resources.
Recommendations

1. Counseling about health insurance issues before patients with CHD leave their parents’ policy or lose their eligibility for children’s services, including information regarding the relationship between education/vocational choices and access to insurance benefits, may be beneficial to facilitate continued insurance (Class IIb; Level of Evidence C).

2. Discussion of the relationship between education/vocational choices and access to insurance benefits should occur early and guide educational planning (Class I; Level of Evidence C).

End of Life, Mortality, and Advance Directives

Long-term survival is an expectation for most individuals with CHD; however, patients with complex CHD remain at increased risk for medical complications and early mortality. It is not uncommon, however, for adolescents and young adults with CHD to hold the misperception that they have been “cured” or “fixed.”178–180 For some patients, the transition period might represent the first occasion when they are confronted with mortality information. Furthermore, patient expectations are often inconsistent with data on life expectancy among patients with CHD. Reid and colleagues47 studied the self-reported life expectancies of 296 patients with moderate to complex CHD between the ages of 16 and 20 years. Patients with CHD expected to live to 75 years of age, only 4 years less than they expected their peers to live; >85% of the patients expected to live much longer than was estimated by the study authors.47 If discussions about life expectancy with healthcare providers were routine, developmentally appropriate, and incorporated as part of general discussion about psychosocial issues, it should help young people have more realistic expectations about their future.

When an adolescent confronts end-of-life issues, the notion of a life cut short can be distressing for patients, their families, and healthcare providers. Freyer noted that for teenagers facing death, “adolescence is a paradox of emerging capabilities and diminishing possibilities.”181 Although it is often difficult for healthcare providers to initiate end-of-life discussions, research suggests that adolescents are often prepared and willing to discuss these issues.182–184 Health professionals should consider several factors when discussing death and end-of-life issues with a young person, including the patient’s developmental level and decision-making capacity, the patient’s conceptualization of death (including personal values), the family’s understanding of death (including religious and cultural beliefs), the patient’s and family’s usual ways of coping, and expectations for the actual death.185–187 These discussions need to be a routine part of care, because patients’ thinking about death is likely to change markedly during the developmental period of adolescence and early adulthood.

Adolescents should be involved in decision-making about palliative and end-of-life care to an extent that is commensurate with their developmental level and prior experience with medical decision making.181,188 Healthcare professionals should consult the laws of their individual states or provinces to determine the specific situations in which adolescents are permitted to provide autonomous consent for medical decisions. Advance care documents allow patients to specify their preferences for medical and palliative care and to identify a legal healthcare decision maker. As such, they allow for the wishes of an individual to be heard and respected when that individual is no longer able to communicate these wishes. Although adolescents’ advance directives might not be legally binding, their preparation can assist with the determination of the adolescent’s preferences and best interests. The process should include patient education about the medical condition, testing, and treatment; clinical determination of the patient’s comprehension of the situation; and solicitation of the patient’s preferences about proposed care.188 McCabe and colleagues185 present a comprehensive list of questions for the assessment of adolescents and families to guide the process of effectively involving adolescents in medical decision making.

The American Academy of Pediatrics advised that “as children develop, they should gradually become the primary guardians of personal health and the primary partners in medical decision-making, assuming responsibility from their parents.”188 Adolescents will benefit from additional decision-making support as they manage end-of-life issues. End-of-life decision-making can be particularly difficult for adolescents, who typically have limited experience in making choices of such magnitude. If the wishes of the adolescent conflict with parental preference, the adolescent may require extra assistance and counseling, particularly if the parents have a history of being overly protective. Although transition is characterized by a process of increased self-determination, the ongoing support and participation of parents when confronting difficult decisions must not be ignored. Many adolescents and young adults will have siblings who would be affected by their death. It is critical to attend to the emotional and practical needs of the entire family during the illness period, as well as after death. It is also important to start to address medical power of attorney and long-term care plans with families of patient with disabilities.

Special Issues for Patients With Developmental Delay/Disabilities

As healthcare providers for adolescents and adults with CHD, we often approach our patients with cognitive impairment and developmental delay differently from those without obvious disabilities. The assumption is that they are unable to successfully transition to adult medical services because of developmental delay, cognitive impairment, or lack of ability to independently manage their own health care. Many pediatric cardiologists have continued to care for adolescents and adults with developmental disabilities. A successful model of healthcare transition for adolescents and adults with CHD must include provisions for patients with developmental disabilities and cognitive impairment.4

The goal of transition in health care for adolescents and adults with special healthcare needs is to maximize lifelong functioning and potential through the provision of high-
quality, developmentally appropriate healthcare services that continue uninterrupted as the individual moves from adolescence to adulthood. To develop appropriate care for patients with developmental disabilities and CHD, we must be committed to creating individualized transition plans, be flexible when necessary, and utilize all available resources to achieve successful transition.

**Practical Implications**

Although the transitional goal for most adolescents and young adults is to eventually accept responsibility for their own health care, this goal may not be realistic for some developmentally delayed patients. ACHD healthcare providers, in conjunction with the primary care physician, must be willing to work with families, vocational counselors, and group home supervisors in situations in which an individual is not able to live and attend to their healthcare needs independently.

The Individuals with Disabilities Act amendment of 1997 mandated that transition planning would be a mandatory part of the annual individual education plan for individuals receiving special education services beginning at age 14 years and continuing through graduation from high school. Adolescents with developmental disabilities and CHD should receive anticipatory guidance at each clinic appointment during adolescence in a manner appropriate for the adolescent’s cognitive ability and level of function. Ideally, ACHD care should be a part of the overall healthcare transition plan, which considers issues such as transportation, community resources, and the individual’s capacity for self-care. A social worker who is familiar with the guidelines provided by state and federal programs for individuals with developmental disabilities can assist in the development of the healthcare transition plan.

ACHD healthcare providers should be familiar with genetic disorders that are commonly associated with both congenital heart defects and developmental delay. Down syndrome, Williams syndrome, and 22q11 deletion disorders (also known as DiGeorge or velocardiofacial syndrome) are the most common ones associated with moderately complex congenital heart defects. Developmental delay, learning difficulties, mental retardation, and other multisystem medical problems are also often present. The American Academy of Pediatrics published health assessment guidelines for children with Down syndrome in 2001 that remain relevant to the adult population. They provide straightforward recommendations on the frequency of medical surveillance for associated health issues at various ages (Table 17). The ACHD healthcare provider should be familiar with these guidelines because the additional health issues associated with genetic disorders can impact the health of their patients and may interact with their patients’ congenital heart disorders. For example, obstructive sleep apnea in an obese adult with Down syndrome and an un repaired ventricular septal defect may lead to worsening hypoxia, pulmonary hypertension, and decompensated polycythemia. Obesity can increase the risk for acquired heart disease, diabetes mellitus, and hypertension.

Guidelines for healthcare supervision of children with Down syndrome and Williams syndrome can be accessed through the American Academy of Pediatrics Web site at http://aappolicy.aappublications.org. Adolescents with 22q11 deletion syndromes often have learning disabilities, developmental delay, and difficulty with speech secondary to a velopharyngeal incompetence, cleft palate, and hearing loss. It is important that patients with CHD, developmental delay, and associated clinical features suggestive of DiGeorge or velocardiofacial syndrome undergo fluorescent in situ hybridization analysis to confirm the diagnosis and for future genetic and reproductive counseling.

**The Transition Clinic**

**Key Elements of the Transition Process**

Regardless of the model of the transition program, there are fundamental principles of transition that have achieved nearly universal endorsement. These principles provide a framework for both individual programs and institutions whose goal is to improve the transition experience for young adult patients with chronic health conditions. These principles are well described in the context of a model that incorporates 3 phases of the transition process: Pretransition, transition, and transfer.

**Pretransition**

Although a formal transition curriculum is most appropriately initiated during adolescence, the concept of transition should be introduced much earlier and continued throughout childhood. As youngsters, patients must be counseled that although they may have a normal or near-normal life, they do not have a normal heart, and they will require lifelong surveillance. Furthermore, they must be taught that CHD is vastly different from other forms of heart disease, which makes it imperative that they follow up with providers specializing in ACHD.

The transition process is a family process; the family plays a critical role that must be supported and encouraged. In this pretransition phase, families should have the expectation that their children will become independent in managing their own medical care by young adulthood. Families should be encouraged to help their children reach this goal. Ideally, using such a strategy of early preparation, the patient and family will enter the transition period with an understand-

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### Table 17. American Academy of Pediatrics Recommendation for Patients With Trisomy 21

<table>
<thead>
<tr>
<th>Annual physical examinations and follow-up beyond adolescence include:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete blood count</td>
</tr>
<tr>
<td>Thyroid function tests</td>
</tr>
<tr>
<td>Audiologic and ophthalmologic evaluation</td>
</tr>
<tr>
<td>Assessment for symptoms of snoring or obstructive apnea</td>
</tr>
<tr>
<td>Assessment of skin problems</td>
</tr>
<tr>
<td>Anticipatory guidance issues to address at each clinic appointment include:</td>
</tr>
<tr>
<td>Diet and exercise issues to maintain appropriate weight</td>
</tr>
<tr>
<td>School and vocational/work issues</td>
</tr>
<tr>
<td>Issues related to group home setting or independent living issues</td>
</tr>
<tr>
<td>Issues related to family relationships, financial planning, and guardianship</td>
</tr>
</tbody>
</table>

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Sable et al. *Best Transition Practices for Adolescents With CHD* 1477
ing of the patient’s heart disease and need for lifelong specialized care.

One model for pretransition counseling uses the transition program advanced practice nurse or physician assistant in this educational role. This practitioner can meet the youngster at ≥1 visits in the pediatric cardiology clinic. During these visits, this practitioner can introduce the concept of transition and transfer, discuss diet and exercise, address high-risk behaviors, discuss contraception and pregnancy, and discuss future educational and vocational goals. This same practitioner can provide continuity of care by following the young adult through the formal transition program and ultimately into the ACHD program. An alternative model uses the pediatric cardiologist in this educational role. In addition, during this pretransition phase, the transition team can coordinate preteen and parent workshops to introduce the concepts of transition and provide a forum in which families can share their fears and concerns with the process.

Transition

When patients are developmentally mature enough, they should enter the formal transition program and complete a transition curriculum. This transition curriculum should ensure that these young people understand their diagnosis and medical history. Ideally, they must understand the normal heart (basic anatomy and physiology) and how the hearts they were born with differ from normal. They can begin to understand the risks of residual hemodynamic burdens and arrhythmic complications. They should be taught to recognize important and concerning signs and symptoms. The transitioning adolescent should have a good understanding of the rationale for previous therapies and options for future medical, surgical, and catheter-based therapies. Finally, they need to be taught to navigate the adult healthcare system; how to access an adult congenital heart specialist; how often they need follow-up; how to access routine health care; how to access emergency health care; and how to navigate the insurance process.

A standard core educational curriculum (Table 18) is an important component of an organized transition process. This education allows patients to gain control of their health and serve as their own advocates. Completing the goals of this curriculum can be accomplished by a variety of educational modalities and should be tailored to individual patients. The curriculum should be of appropriate breadth and depth but concise enough to be completed before patients move into the ACHD clinic. This may require 2 clinic visits for mature, well-adjusted patients with mild disease, mild functional limitations, and a strong support system or many visits with intense peer support-group involvement for a young person with learning disabilities, complex disease, significant functional limitations, and no support system. Flexibility is a key to success.

The example of a transition curriculum outlined in Table 18 includes residual hemodynamic and arrhythmia considerations, noncardiac surgical and medical problems, contraception and pregnancy planning, vocational and insurance planning, and other lifestyle issues. The curriculum goals can be accomplished in a number of ways. Much of the curriculum can be addressed at routinely scheduled clinic visits with the participation of physicians, advanced practice nurses or physician assistants, and other support services. In addition, introductory seminars for patients and families, peer support groups, and informational seminars can be used to address some of the components of the curriculum.

### Table 18. Transition Curriculum Topics

<table>
<thead>
<tr>
<th>Residual hemodynamic considerations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemodynamic issues</td>
</tr>
<tr>
<td>Symptoms and how to respond</td>
</tr>
<tr>
<td>Diagnostic tools in follow-up</td>
</tr>
<tr>
<td>Management options</td>
</tr>
<tr>
<td>Arrhythmia considerations</td>
</tr>
<tr>
<td>Risks</td>
</tr>
<tr>
<td>Signs and symptoms</td>
</tr>
<tr>
<td>Screening tools</td>
</tr>
<tr>
<td>Diagnostic tools</td>
</tr>
<tr>
<td>Management options</td>
</tr>
<tr>
<td>Endocarditis considerations</td>
</tr>
<tr>
<td>Risks, implications, recognition, and response</td>
</tr>
<tr>
<td>Prevention</td>
</tr>
<tr>
<td>Contraception and pregnancy planning</td>
</tr>
<tr>
<td>Contraceptive options and risks</td>
</tr>
<tr>
<td>Risks of pregnancy to mother and to fetus</td>
</tr>
<tr>
<td>Management of pregnancy plan</td>
</tr>
<tr>
<td>Noncardiac surgery considerations</td>
</tr>
<tr>
<td>Risks</td>
</tr>
<tr>
<td>Location of surgery</td>
</tr>
<tr>
<td>Knowledge and skills of surgical team</td>
</tr>
<tr>
<td>Noncardiac medical problems</td>
</tr>
<tr>
<td>Access to appropriate care</td>
</tr>
<tr>
<td>Career, vocational, and insurance planning</td>
</tr>
<tr>
<td>Lifestyle issues</td>
</tr>
<tr>
<td>Marriage and family planning</td>
</tr>
<tr>
<td>Education</td>
</tr>
<tr>
<td>Employment</td>
</tr>
<tr>
<td>Life and health insurance</td>
</tr>
<tr>
<td>Learning disabilities</td>
</tr>
<tr>
<td>Anxiety and depression</td>
</tr>
<tr>
<td>High-risk behaviors</td>
</tr>
<tr>
<td>Healthy eating</td>
</tr>
<tr>
<td>Physical fitness</td>
</tr>
<tr>
<td>Salt and fluid restriction (if warranted)</td>
</tr>
<tr>
<td>Relative safety of exercise and hobbies</td>
</tr>
<tr>
<td>End-of-life decisions</td>
</tr>
<tr>
<td>Skills training</td>
</tr>
<tr>
<td>Communication</td>
</tr>
<tr>
<td>Decision making</td>
</tr>
<tr>
<td>Creative problem solving</td>
</tr>
<tr>
<td>Assertiveness</td>
</tr>
<tr>
<td>Self-care</td>
</tr>
<tr>
<td>Self-advocacy</td>
</tr>
</tbody>
</table>
Transfer

Timing of Transfer

Ideally, transfer of care from the pediatric to adult healthcare system occurs at the successful completion of a thoughtful transition process. When deciding on the timing of transfer, 2 important points need to be considered: There should be a policy on timing, and this policy should be flexible.

First, there should be an institutional policy on timing of transition to ensure that transition and transfer actually occur, and in a predictable manner. When patients and families have an explicit target age, they recognize that transition and transfer will occur, appreciate their active involvement in the process, and are prepared for the ultimate transfer of care. Additionally, when transition is the “rule,” young people see the process not as something they as individuals are forced to go through alone but rather as a natural process that everyone goes through; much as high school follows junior high, so adult care follows pediatric care. Finally, a policy on timing ensures that all patients have the opportunity to take full advantage of the transition curriculum.

Second, a transition program must be flexible with respect to timing and should tailor the process to the developmental and psychosocial status of each young person. Transition should be completed only after the young patient has accomplished the developmental tasks of adolescence and has demonstrated the ability to manage his or her health care independent of his or her family and pediatric provider.

Adolescents have many difficult tasks to accomplish before they leave this stage in development. They must begin to develop a personal sense of identity, which normally occurs in young adulthood; adjust to this new physical sense of self; adjust to new intellectual abilities and the increased cognitive demands placed on them at school and in society; develop educational and vocational goals and a plan to meet these goals; establish emotional and psychological independence from their parents and begin to adopt a personal value system; learn to manage their sexuality and begin to develop intimate relationships; and develop increased impulse control and behavioral maturity. It is important to recognize that chronic illness, physical disability, and cognitive limitations can disrupt the usual developmental trajectories. This makes the accomplishment of such tasks more difficult and thus the process of transition more challenging.

Furthermore, the sometimes overprotective attitudes of pediatric providers and patient families can diminish self-esteem and hinder the development of self-sufficiency. In general, the adult healthcare system demands a higher level of personal responsibility and autonomy than the pediatric system. Thus, to be effective healthcare consumers in the adult healthcare environment, young people should demonstrate the ability to meet their healthcare needs independent from their families and pediatric providers before transfer; make their own appointments, meet independently with their healthcare providers; administer their own medications and other treatments; understand their medical history; and recognize signs of clinical deterioration.

Finally, transfer of care during medical crises or periods of psychosocial disequilibrium should be avoided. All too often, patients are transferred when they become pregnant, mentally ill, or noncompliant, or after they experience their first “adult” complication. Transfer during these times precludes maximal utilization of the transition curriculum and imposes a tremendous psychological burden on the patient.

Adult Provider Services

Although the needs of some patients are relatively straightforward, others have complex medical needs and may require the involvement of a variety of subspecialty consultants, in addition to capable primary healthcare providers willing to coordinate and manage the complex care they need. Other medical and nonmedical professionals critical to successful transfer include advanced practice nurses or physician assistants, social workers, and those who specialize in vocational and educational issues.

Coordinated Transfer Process

A coordinated transfer process is the final component of a successful transfer process. A carefully prepared health summary allows seamless transfer of care and provides a blueprint for the new healthcare team. It should comprise a complete medical history that includes diagnoses and previous interventions, a medication list, laboratory values, and other diagnostic studies, as well as information about the patient’s functional status, the tempo of disease progression, and the impact of other comorbidities. Information about psychosocial concerns, end-of-life preferences, the extent of family involvement, and adherence issues should also be communicated if available. A comprehensive summary avoids the need to reinvent the therapeutic wheel and helps to prevent errors being made or mistakes being repeated. In addition to the formal medical summary, it is vital that the pediatric providers communicate directly with their counterparts in the adult healthcare system. The more integrated pediatric and adult providers are, the more seamless the transition process will be.

It is important that the adult providers respect the therapeutic plan that was established by the pediatric providers and communicated to the young patients and their families. As explained above, the immediate reevaluation and drastic change in management that many new adult providers are tempted to make after care transfer can often be overwhelming to patients and diminish their trust. Regardless, one must also take advantage of the unique opportunity this transition process provides to take a fresh look at the patients as evolving adults and to reassess therapeutic options in the light of new technologies. This process can be done in conjunction with the pediatric providers and after a period of introduction and relationship development in the transition process. In this way, trust is not lost, and the best possible plan for the future is made.

Many transition “tools” have been suggested to aid in a smooth transition and transfer. For example, a central provider, such as an advanced practice nurse or physician assistant, who can assume responsibility for the entire process has been shown to be helpful. The coordinator can share membership on both the pediatric and adult teams and serve as a liaison between them, as well as serving as a reassuring presence and advocate for patients and families. In addition, to address the inevitable
uncertainty felt by young people and their families about impending transfer, many programs have created a transfer “package” with information about the adult programs. Finally, patients who have already transferred can play a part in welcoming young people who are transferring their care.

Because increasing numbers of young people with complex congenital illnesses are surviving into adulthood, there is an urgent need for programs designed to facilitate their smooth movement from pediatric to adult healthcare environments. It is hoped that in the near future, transition programs will become the standard of care, making it more likely that patients with complex chronic illness can achieve their full potential under appropriate medical surveillance and live meaningful and productive lives.

Disclosures

Writing Group Disclosures

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<tr>
<th>Writing Group Member</th>
<th>Employment</th>
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This table represents the relationships of writing group members that may be perceived as actual or reasonably perceived conflicts of interest as reported on the Disclosure Questionnaire, which all members of the writing group are required to complete and submit. A relationship is considered to be “significant” if (1) the person receives $10,000 or more during any 12-month period, or 5% or more of the person’s gross income; or (2) the person owns 5% or more of the voting stock or share of the entity, or owns $10,000 or more of the fair market value of the entity. A relationship is considered to be “modest” if it is less than “significant” under the preceding definition.

*Modest.
†Significant.
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