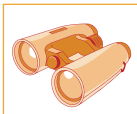

Primary and General Care of the Child with Congenital Heart Disease

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For children with congenital heart disease (CHD), primary care needs and cardiology concerns frequently overlap, with different issues coming to the forefront at different ages (Table 1). This review will focus on a few key points for each age group.

Growth and Nutrition

Congenital heart disease is frequently associated with malnutrition and growth retardation. As many as 27% of children with CHD fall below the third percentile for height and weight. The degree of growth impairment does not always correlate with the severity of the cardiac lesion. Even among children with relatively mild hemodynamic disturbances, a disproportionately large number grow below the tenth percentiles. There are multiple causes of growth failure (Table 2). Some factors, such as hemodynamic disturbances and higher basal metabolic rates, can and should be aggressively addressed. Other factors, such as intrauterine growth retardation and genetic syndromes, can be identified but usually cannot be altered. Since optimal outcome depends on maximizing weight gain and linear growth, close attention to nutritional status is crucial for the care of all children with CHD.

Different patterns of growth impairment are observed in different types of CHD. Cyanotic patients have disturbances in both height and weight, while acyanotic patients tend to have more problems with weight gain than with linear growth. With left to right shunts, the degree of growth impairment is proportional to the size of the shunt and the severity of the pulmonary hypertension. Failure to thrive is most pronounced in cyanotic patients with congestive heart failure (CHF), but growth impairment does not correlate well with the degree of hypoxemia. In cyanotic patients, iron deficiency has been associated with stroke and behavioral problems as well as poor growth and may be masked by polycythemia. Patients with pressure overload lesions without intracardiac shunts grow normally.

Both increased metabolic demands and decreased caloric intake contribute to growth failure in children with CHD. Like other malnourished children, infants with CHD with growth retardation have a higher basal metabolic rate (BMR). This is primarily due to an increased proportion of metabolically active tissue and decreased fat stores; their metabolic rate is normal when adjusted per kilogram of lean body tissue. Infants with CHD need significantly higher daily caloric intakes than recommended for average children, often up to 160 kcal/kg/day. Compounding this problem, these infants typically take in fewer calories than needed even for normal growth. Tachypnea, increased work of breathing, diminished strength and easy fatigability, as well as difficulty with coordination of sucking and swallowing all contribute to inadequate oral intake. Infants with CHD also exhibit specific gastrointestinal problems such as delayed gastic emptying, impaired gastrointestinal motility, and decreased gastric capacity. They do not demonstrate a consistent pattern of malabsorption however.

Other factors may affect growth even in the face of adequate caloric supplementation. There is a higher incidence (6–14%) of intrauterine growth retardation among children born with cardiac lesions even when neonates with extracardiac anomalies are excluded. Many genetic syndromes associated with congenital heart disease alter growth. Growth impairment is a part of these syndromes but might also warrant evaluation of endocrine status for hormonal deficiencies. Similarly, poor growth in a child with mild cardiac disease should prompt further investigation and might lead to the diagnosis of a previously unrecognized genetic syndrome. At times, it may be difficult to determine whether poor weight gain is secondary to the hemodynamic disturbance or to the underlying genetic predisposition. In these situations, early cardiac repair to eliminate at least one source of growth failure may be reasonable.

Regular assessment of growth parameters, including plotting growth curves, is very important. Evaluation should include assessment of the adequacy of caloric intake and possible need for supplemental tube feedings. Increased energy intake may be accomplished with fortification of feedings (Table 3). If increases beyond 24 kcal/oz are needed, as for patients with severe fluid restrictions, then supplementation with fat products or low-osmolality glucose polymers is preferable to avoid excessive renal solute load and osmolality. Breast milk remains the best source of nutrition for infants with CHD; however, it should be fortified to meet their increased metabolic needs. Despite increased caloric density, some infants may not grow adequately with oral feedings alone. Infants with CHD receiving 24-hour continuous NG feedings achieve desired intakes and improved growth most consistently. To balance this beneficial effect with the risk of oral-motor dysfunction, infants may be allowed to take calorically dense oral feeds throughout the day and then be given continuous NG feeds overnight.

Most infants with CHD can grow with these nutritional interventions. Failure to achieve adequate weight gain despite maximal enteral supplementation may be evidence of significant hemodynamic compromise and thereby may be an indication for surgery. In the immediate post-operative period, early nutritional support improves outcomes. Sig-

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Table 1. Age of Presentation of Specific Primary Care Problems in Patients with Congenital Heart Disease

At diagnosis
Prognostic implications of specific diagnosis
Psychological impact on the family
Medications—benefits, side effects, compliance
Screen for associated extracardiac malformations
Social support mechanisms
Financial assistance
0-2 Years
Growth
Nutrition
Development
Immunizations
3-6 years
Endocarditis prophylaxis
Dental care
Special dietary recommendations
Occult infections
6-12 years
School program
Activity recommendations
Endocarditis prophylaxis
Scoliosis detection
Occult infections
Heart-healthy diet
Adolescence
Medications-compliance
Athletics
Risk-taking behavior
Delayed puberty
Fears concerning sexual performance
Contraception, pregnancy, genetic counselling
Heart healthy diet
Endocarditis prophylaxis
Vocational counseling
Occult infections
Young adulthood
Shared issues with adolescent list
Employability
Insurability
Preventive counseling
Transition to adult care team
Advance directives
At death of patient
Assess grief reaction and support
Provide genetic counselling
Discuss other family concerns
Establish follow-up appointment

nificant catch-up growth is possible after repair. In general, more catch-up growth occurs when surgery is performed at less than 2 years of age rather than later and after corrective surgery more than after palliation. Typically weight picks up more quickly than linear growth. When surgical repair does not result in substantial catch-up growth, the patient should be evaluated for significant residual hemodynamic defects and for complications of surgery, although continued growth impairment may be secondary to non-cardiac factors.

Table 2. Causes of Growth Retardation in Children with CHD

Genetic/Extracardiac	Increased Metabolic Demands	Insufficient Nutrient Intake
Intrauterine growth retardation	Increased basal metabolic rate	Fatigability/weakness
Genetic syndromes	Hemodynamic abnormalities	Tachypnea
Extracardiac anomalies	Congestive heart failure	Poor oral feeding skills
Hormonal deficiencies	Chronic hypoxemia	Iron deficiency

Table 3. Increasing Caloric Density of Feedings

Human Milk Fortifier (Enfamil, Mead Johnson), 1 packet per 25 mL of BrM = 24 kcal/oz
Formula concentration to 24 kcal/oz:
1 cup powdered formula + 3 cups water or
4 oz ready-to-feed + 1/2 scoop powdered formula
Supplementation to 26-30 kcal/oz:
Fat modular products
Medium chain triglycerides (MCT) oil (Mead Johnson), 8 kcal/mL
Microlipid (safflower oil emulsion, Mead Johnson), 4.5 kcal/mL
Low-osmolality glucose polymers
Polycose (Ross), 23 kcal/tablespoon
Moducal (Mead Johnson), 30 kcal/tablespoon
Pediasure (Ross), 30 kcal/oz ready-to-feed (for children over 1 year of age)

Many children with CHD grow normally and need no special dietary advice. On the other hand, some children with CHD develop a sedentary lifestyle, either because of imposed activity restrictions or because of family overprotectiveness. In these children, attention should be paid to counseling about "heart healthy" eating with a diet low in saturated fat and cholesterol. Obesity, deconditioning and hypertension complicate the assessment of cardiovascular symptoms and negatively affect long-term outcome. Preventive cardiology counseling should be a routine part of care.

Immunizations

With few exceptions, children with congenital heart disease should receive routine immunizations according to the schedule published by the American Academy of Pediatrics (available in the Red Book or at http://www.aap.org/). Infants with CHD are at increased risk of immunization delay due to missed or postponed immunizations during hospitalizations or illnesses. Consideration should be given to vaccination prior to discharge for all infants with delinquent immunizations. Post-operative status per se is not a contraindication to immunization. Some cardiologists are reticent to give immunizations in patients for whom fever would complicate care. In the past, most such febrile reactions were following administration of vaccines containing whole cell pertussis (DTP). With the acellular pertussis formulation (DTaP) now in use, the risk of fever is substantially lower. These considerations must be weighed along with the fact that *B. pertussis*, *H. influenzae B*, and *S. pneumoniae* still are not uncommon pathogens and can cause life-threatening infections in infants. Inactivated polio vaccine (IPV) has replaced oral polio vaccine (OPV) in the routine schedule and can be administered in hospitalized patients. A protein conjugate pneumococcal vaccine, im-

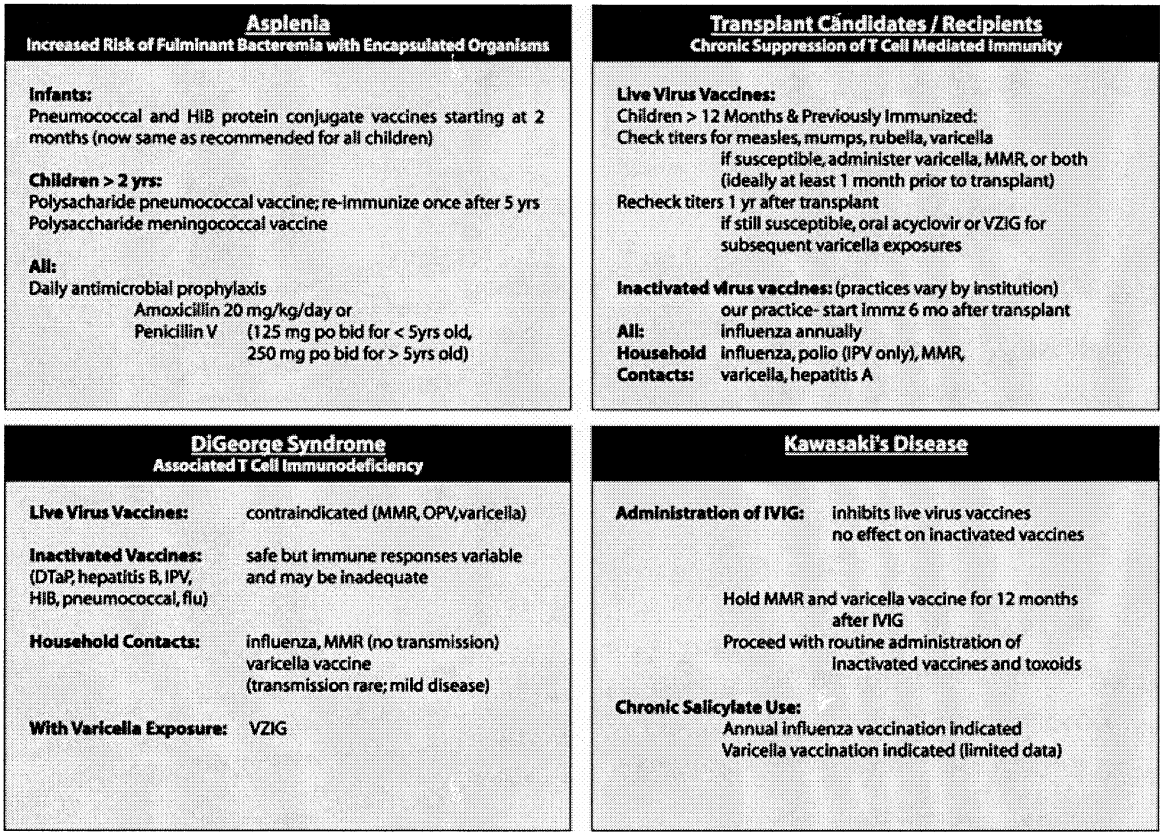

Figure 1. Immunization issues in subsets of patients with congenital heart disease.

munogenic in infants under 2 years of age, recently has been added to the routine immunization schedule.

Patients with asplenia, DiGeorge syndrome, cardiac transplantation and Kawasaki's disease have special immunization needs (Figure 1). Certain vaccinations also deserve special attention in patients with CHD. Yearly influenza immunization each autumn is recommended for all children 6 months of age or older with hemodynamically significant cardiac disease. It also is recommended for children requiring long-term aspirin therapy. Household contacts of young infants with significant CHD should be immunized. Children who are electively scheduled for cardiac interventions should be immunized against hepatitis B to protect against transfusion-related infection. In particular, this may be an issue for school-age children who received their other immunizations before the recommendations for universal neonatal hepatitis B vaccination and for adolescents who are less likely to complete the series. Children receiving long-term salicylate therapy are at increased risk of Reye's syndrome with wild-type varicella infection and thus theoretically would benefit from vaccination. However it is unknown whether Reye's syndrome occurs after varicella vaccination with concomitant salicylate use. Although no cases have been reported, the manufacturer recommends that salicylates not be administered for 6 weeks after the varicella vaccine. In the absence of data, clinicians must weigh the theoretical risks of vaccination against the known risks of wild-type infection and the risks of interruption of aspirin therapy for each individual patient.

Prevention of respiratory synctial virus (RSV) infection remains debatable. RSV-IGIV is contraindicated in patients with unoperated cyanotic congenital heart disease secondary to adverse outcomes in the initial trials. Patients with asymptomatic acyanotic heart disease with appropriate respiratory indications may benefit from prophylaxis. There is an ongoing multicenter clinical trial evaluating the safety and efficacy of palivizumab, a monoclonal antibody administered intramuscularly, in patients with CHD; preliminary results are not yet available.

SBE Prophylaxis

Antimicrobial prophylaxis against bacterial endocarditis is important for nearly all patients with CHD. Reinforcement of the need for SBE prophylaxis should occur at each visit. Detailed recommendations are easily accessible online (http://www.americanheart.org).

School and Activities

Many factors affect development and school performance in children with CHD. Chronic hypoxemia adversely affects development, particularly gross motor development. In cyanotic patients, surgery improves intelligence quotient

(IQ). In this group, older age at repair negatively correlates with intellectual and gross motor performance. Longstanding congestive heart failure also causes developmental delay. Associated genetic syndromes may influence both intelligence and motor skills. Furthermore, complications of interventions, such as perioperative stroke, may result in specific deficits. Discussion of the effects of cardiopulmonary bypass and circulatory arrest on neurodevelopmental outcome is beyond the scope of this review. However, recent studies are reassuring that most children who undergo cardiac surgery do quite well. Even patients who have undergone the Fontan procedure for palliation of single-ventricle physiology generally score in the normal range on intelligence tests in the preschool and early school years. In individual patients, though, even simpler surgeries, such as atrial septal defect repair, may result in subtle neurologic abnormalites. Overall there is a broad range of IQ among patients with CHD. The presence of heart disease certainly does not preclude excellent academic performance.

Direct communication with teachers, principals, and school nurses helps to educate those directly involved about relevant health issues. For example, patients who fatigue easily may have better school performance if their classes are clustered close together to avoid their having to walk further or climb stairs. Specific information may also alleviate unspoken fears of the staff. At times it may be useful to have open classroom discussions, such as demonstrations of how pacemakers work, to facilitate student acceptance of and tolerance for the patient's special needs. For other patients, emphasis may need to be placed instead on the avoidance of an overly protective environment and promotion of optimal social adjustment at school.

The majority of patients with CHD may participate fully in all activities appropriate for their age, interests and abilities. A letter stating this specifically may be needed for certain activities and athletics. Most other patients can be encouraged to take part in recreational activities but should be allowed to pace themselves and rest when necessary. Children who have hemodynamically significant heart disease are discouraged from participating in competitive sports because of the likelihood of their overexerting themselves due to perceived pressure from teammates or coaches. Specific contraindications to maximal exercise include exercise-induced arrhythmias, cardiomyopathy, pulmonary artery hypertension and evidence for myocardial ischemia at rest or with exercise. Following reparative operations most children may participate in competitive sports.

Adolescents and Young Adults with CHD

Young adults with congenital heart disease are becoming an increasingly important part of practice for both adult and pediatric cardiologists. Certain issues warrant special attention. Adolescents are at particular risk of non-compliance. Although difficult in all patients, non-compliance may have life-threatening consequences in patients requiring immunosuppression after transplant or needing anti-coagulation. Additionally, adolescents who have lived with serious chronic illnesses exhibit significantly more risk-taking behavior than other teens. Employability and insurability may be difficult problems for young adults who are symptomatic and may be barriers to ongoing medical care. Of note though, in long-term natural history studies of patients with CHD, more patients graduated from college than controls, and patients were employed at the same rate as the general population.

Fears about sexual performance as well as issues about pregnancy and contraception should be addressed directly. Counseling about pregnancy prevention in young women with high-risk heart disease should begin early in adolescence. Parenterally administered or implanted contraceptive agents, such as medroxyprogesterone acetate and levonorgestrel, are preferred methods of contraception in these patients because of their high efficacy and long duration of action. Low estrogen oral contraceptives are effective and safe except in women with right-to-left shunting, polycythemia or a prior thromboembolic event. Abstinence and barrier methods have lower efficacy, and intrauterine devices are not recommended because of infection risk. In general, assessment of the severity of the patient's heart disease and risk associated with pregnancy for both the mother and the fetus should occur prior to childbearing age so that interventions that might decrease these risks may be performed prior to conception. Management is much more difficult when the patient presents pregnant and then is found to have significant heart disease. Severe systemic ventricular failure, right ventricular hypertension with 50–70% systemic pressures, oxygen saturations less than 82% and hemoglobin greater than 16 g/dL are predictors of high morbidity and mortality for both mother and fetus. Genetic counseling is specific to the lesion and family history. Recurrence risk is highest for patients with left-sided obstructive lesions and for patients with associated chromosomal abnormalities. Fetal echocardiography is indicated. Although many common cardiac drugs are safe during pregnancy, angiotensin-converting enzyme inhibitors, amiodarone and warfarin are contraindicated. Close coordination between the obstetrician and the cardiologist, with attention to disease-specific principles of management, optimizes outcome and will allow the majority of women with CHD to have safe pregnancies, deliveries and post-partum recovery.

Care of the young adult with CHD requires familiarity with congenital heart lesions, their natural history in the repaired and unrepaired state, historical knowledge of prior surgical approaches and awareness of potential long-term complications from corrective or palliative procedures. Cardiac issues must be incorporated into the general context of the patient's adult medical needs. How this is best accomplished varies by institution and geographic location but ideally a team approach is used. Most importantly,

patients who have been followed regularly throughout childhood should continue to receive the same level of care into adulthood with access to well-trained and interested specialists.

Summary

Patients with significant CHD have special primary care needs that vary by age. A comprehensive approach with communication and coordination between the primary care physician, cardiologist, other specialists, state programs, school and parents optimizes both cardiac and general care of these patients.

Suggested Reading

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