Congenital Heart Disease Guidelines Target Adults

BY DIANA MAHONEY
New England Bureau

The unique lifetime care needs of adults with congenital heart disease—particularly young adults who are making the transition out of pediatric cardiology care—are the focus of new practice guidelines released jointly by the American College of Cardiology and the American Heart Association.

The adult congenital heart disease (CHD) guidelines, the first of their kind in the United States, are designed to help physicians make routine clinical decisions for the ever-increasing number of patients who, because of the advances in the treatment of CHD, are now surviving into adulthood but are doing so with complex cardiac anatomy and physiology that are related to the repaired heart defects, according to Dr. Carole A. Warnes and her colleagues on the American College of Cardiology/American Heart Association guidelines writing committee.

"In particular, there are a substantial number of young adults with single-ventricle physiology, systemic right ventricles, or complex intracardiac baffles who are now entering adult life," the authors wrote in the executive summary of the guidelines, which will be published in the Dec. 2, 2008, issues of both the Journal of the American College of Cardiology and Circulation and which are available in the online editions of each issue (doi:10.1016/j.jacc.2008.10.001).

Altough the infrastructure of most pediatric cardiology centers supports the multiple unique needs of children with CHD through, for example, comprehensive case management by advanced practice nurses and social workers, the adult health care system is not similarly structured, the authors noted.

In addition, "young adults have many psychological, social, and financial issues that present barriers to proactive health management," they wrote. In an effort to minimize "the compound effects of a complex and unfamiliar disease with an unprepared patient and healthcare system," the practice guidelines outline the most important diagnostic and management strategies and indicate when to refer patients to a highly specialized center.

Improving the delivery of care and ensuring access to care figure prominently in the recommendations. Specifically, the guidelines call for the development and strengthening of transition clinics for adolescents and young adults with CHD, as well as outreach and education programs for patients, families, and caregivers and enhanced education of adult and pediatric cardiovascular specialists to ensure optimal management of adult CHD.

The guidelines recommend that:

► The care of adult CHD patients should be coordinated by regional centers of excellence.

► Adult CHD patients carry a complete "medical passport" containing their medical histories as well as contact information for the regional centers of excellence to facilitate access to data and counsel.

► Designated health care guardians be included in the medical decision-making process for patients whose care is complicated by additional special needs.

► Patients have a primary care physician and a local cardiovascular specialist, each of whom has copies of current clinical records on file.

► Patients establish a relationship with a regional adult CHD center to ensure the availability of geographically accessible care when needed.

The guidelines specify that, in the absence of specific training or experience in adult CHD, primary caregivers and cardiologists of patients with CHD should work in collaboration with trained specialists, and patients should have access to specialized follow-up care. For example, the guidelines state that patients with low-risk, simple CHD should have at least one follow-up at a regional adult CHD center, while more frequent follow-up (every 12-24 months minimum) is advised for "adults with complex and moderate CHD."

In addition, certain clinical scenarios warrant consultation with, treatment at, or transfer to, a regional adult CHD center. Such scenarios include hospital admission for urgent or acute care in most cases; the performance of diagnostic or interventional procedures; surgical procedures requiring general anesthesia or conscious sedation; urgent or acute care of cardiac problems; and urgent or acute

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care of noncardiac problems in high-risk patients.

The guidelines also address the psychosocial needs of adult CHD patients with the recommendation that the comprehensive care of these patients should incorporate individual and family psychosocial screening, counseling, and education regarding the possible social, emotional, and vocational impact of the conditon.

Because CHD patients are at increased risk for infectious endocarditis, it is important that patients and their families be educated about the signs and symptoms of infectious complications, as well as how to prevent them, according to the authors.

In particular, the guidelines recommend antibiotic prophylaxis in high-risk CHD patients "before dental procedures that involved manipulation of the gingival tissue or the periapical region of teeth or perforation of the oral mucosa." Antibiotic prophylaxis also should be considered before vaginal delivery at the

An increasing number of patients are now surviving into adulthood with complex cardiac anatomy and physiology.

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time of membrane rupture in patients with a prosthetic cardiac valve or in whom prosthetic material was used for valve repair and patients with unrepaired and palliated cyanotic CHD.

However, antibiotic prophylaxis against infectious endocarditis "is not recommended for nondental procedures [such as esophagogastroduodenoscopy or colonoscopy] in the absence of active infection," the authors wrote in the guidelines.

Pregnancy and contraception require special consideration in women with CHD. With respect to contraception, oral estrogen-containing drugs are not recommended for patients at risk of thromboembolism, including those with pulmonary arterial hypertension or cyanosis related to an intracardiac shunt, according to the guidelines. Regarding pregnancy, patients are advised to consult with an adult CHD expert to determine a labor and delivery management plan prior to becoming pregnant.

In addition to the general recommendations for the care of adult CHD patients, the guidelines also include comprehensive information on the clinical features, diagnosis, treatment options, activity limitations, pregnancy risks, and preventive strategies related to specific lesions, such as atrial, ventricular, or atrioventricular septal defects; patent ductus arteriosus; left-sided heart obstructive lesions; right ventricular outflow tract obstruction; pulmonary artery hypertension/Eisenmenger physiology; and tetralogy of Fallot.

The adult CHD guidelines were developed in collaboration with the American Society of Echocardiography, the Canadian Cardiovascular Society, the Heart Rhythm Society, the International Society for Adult Congenital Cardiac Disease, the Society for Cardiovascular Angiography and Interventions, and the Society of Thoracic Surgeons.

While the recommendations are evidence based wherever possible, "unlike other ACC/AHA practice guidelines, there is not a large body of peer-reviewed published evidence to support most recommendations," the authors wrote. For this reason, the evidence supporting many of the recommendations comes from the consensus of experts. ■

In RA Patients, Cardiovascular Risk Matches Type 2 Diabetes

BY BETSY BATES
Los Angeles Bureau

SAN FRANCISCO — Patients who have rheumatoid arthritis should be assessed annually for cardiovascular risk factors, a recommendation necessitated by a heart disease risk profile that equates to that of those with type 2 diabeties, a European task force concluded.

"Cardiovascular risk management is urgently needed for patients with rheumatoid arthritis, ankylosing spondylitis, and psoriatic arthritis," said Dr. Michael T. Nurmohame, who was speaking on behalf of the European League Against Rheumatism cardiovascular disease risk management task force at the annual meeting of the American College of Rheumatology.

Task force recommendations highlighted at the meeting included:

► Characterizing of rheumatoid arthritis, ankylosing spondylitis, and psoriatic arthritis as "high-risk" conditions with regard to cardiovascular disease, similar to diabetes.

► Launching annual screening for cardiovascular risk of every RA patient, with consideration of screening of ankylosing spondylitis and psoriatic arthritis patients as well.

► Providing every patient with lifestyle recommendations for lowering cardiovascular risk.

► Emphasizing aggressive control of disease activity to suppress inflammation and lower cardiovascular risk.

► Adapting cardiovascular risk scoring models (such as the newly adapted Systematic Coronary Risk Evaluation SCORE) by a factor of 1.5 to account for elevated baseline risk associated with inflammatory rheumatic diseases.

► Considering of treatment with statins and/or antihypertensive drugs according to cardiovascular management targets established by local guidelines; or, if no local guidelines exist, when targets exceed 10-year cardiovascular mortality risk models established in the newly adapted SCORE.

► Acknowledging that the role of cyclooxygenase-2 inhibitors and nonsteroidal anti-inflammatory drugs is not well established in RA patients.

► Limiting corticosteroids to the lowest possible doses.

The task force consisted of 21 rheumatologists, internists, cardiologists, and epidemiologists representing nine European countries.

Its work was prompted by the increasing recognition that those patients who have rheumatoid arthritis face a steeply elevated risk in cardiovascular diseases, said Dr. Nurmohamed, who is a rheumatologist at the VU University Medical Center and Jan van Breemen Institute in Amsterdam.

The risk can only be partially explained by traditional risk factors, with inflammatory processes serving as the apparent "missing link," he suggested.

Earlier this year, Dr. Nurmohamed and his associates published the results of the CARRÉ study, in which they compared cardiovascular risk in 353 patients with rheumatoid arthritis with two groups of similarly aged patients who were enrolled in the population-based Hoorn cohort study: 194 of the patients had type 2 diabetes and 258 healthy controls (Ann. Rheum. Dis. 2008 Aug. 12 [doi:10.1136/ard.2008.094151]).

The prevalence of cardiovascular disease was 5% in nondiabetic patients with no rheumatoid arthritis; 12.4% in patients with type 2 diabetes; and 12.9% in patients with RA.

Some of that risk can be accounted for by increased hypertension, dyslipidemia, and lifestyle factors in the RA population, he said.

However, inflammatory rheumatic diseases themselves also seem to confer an independent risk that should be accounted for in models that predict cardiovascular mortality, Dr. Nurmohamed commented. ■

Restrictions on Ranolazine's Label Lifted, Cuts HbA_{1C}

BY ELIZABETH MECHCATIE
Senior Writer

The Food and Drug Administration has approved a revised indication and several label additions for the angina drug ranolazine, including a statement that the drug reduced hemoglobin A_{1c} in people with diabetes.

The indication is still for "the treatment of chronic angina, but "the second-line restriction on the use of ranolazine to treat patients with chronic angina has been removed," according to an announcement issued by the FDA .

Previously, ranolazine's indication was restricted to patients who had not had an adequate response with other antianginal drugs.

Previously, the indication was for treatment of chronic angina, but with the added statement that it should be reserved for patients who have not had an adequate response with other antianginal drugs, because ranolazine increases the QT interval.

The additional statement has been removed from the revised label, with the information about the QT interval prolongation now in the warnings and precaution section.

Also added to the label is a statement that cites the significantly lower rate of arrhythmias in patients with coronary heart disease who were treated with ranolazine in the MERLIN-TIMI 36 trial, compared with those on placebo, CV Therapeutics Inc. noted in its announcement of the approval.

The indications section of the revised label also says that the drug can be used with β-blockers, nitrates, calcium channel blockers, antiplatelet therapy, lipid-lowering therapy, ACE inhibitors, and angiotensin receptor blockers.

CV Therapeutics markets ranolazine in extended-release tablet form as Ranexa, which was approved in January 2006.

Ranolazine has antianginal and anti-ischemic effects, but its exact mechanism of action is not known, according to the label.

In a statement, the company said that data from the MERLIN-TIMI 36 trial were submitted to the FDA in September 2007, as part of its supplemental application. The revised label includes the statement that in the study—which compared ranolazine to placebo in more than 6,000 patients with acute coronary syndrome—no benefit was seen on outcome measures, but that the study was "somewhat reassuring regarding proarrhythmic risks, as ventricular arrhythmias were less common on ranolazine."

The incidence of arrhythmias (ventricular tachycardia, bradycardia, supraventricular tachycardia, and new atrial fibrillation) was 80% among those treated with ranolazine, compared with 87% of those on placebo, a significant difference, according to the label. However, the label also states that the difference in arrhythmias did not result in lower mortality, or reductions in arrhythmia hospitalizations or arrhythmia symptoms.

The label notes that there were no proarrhythmic effects seen on 7-day Holter recordings in 3,162 patients with acute coronary syndrome who were treated with ranolazine.

The revised label also includes the statement that ranolazine "produces small reductions in [hemoglobin A_{1c}] in patients with diabetes, the clinical significance of which is unknown," and that the drug "should not be considered a treatment for diabetes." ■