Technical advances in medical and surgical treatment of patients with congenital heart disease and advances in bacteriology and cardiac imaging have been associated with substantial changes in the clinical profile of infective endocarditis. Endocarditis after congenital heart surgery is a concern because postoperative residue and sequelae are potential foci (1-7). Hemodynamic monitoring with indwelling catheters causes areas of sterile endocardial damage of right-sided cardiac valves, increasing susceptibility to infection during bacteremia.

Cardiac lesions at risk. These lesions are characterized by increased velocity of flow at sites of significant pressure gradients, with the maximal deposition of organisms at the site of the low pressure sink immediately beyond the orifice or at the site of jet impact. Tetralogy of Fallot, physiologically analogous forms of cyanotic congenital heart disease and congenital aortic valve disease account for approximately 35% of reported cases of infective endocarditis (1-7). Pulmonary valve stenosis is reported less frequently, and in correlation of the aorta, the principal site of infective endocarditis is on a coexisting bicuspid aortic valve. Other frequently involved lesions include restrictive ventricular septal defects and patent ductus arteriosus. The left anterior leaflet with mitral regurgitation in patients with ostium primum atrial septal defect represents a risk, but uncomplicated ostium secundum atrial septal defect is at little or no risk. Postoperative infective endocarditis in congenital heart disease is most frequently reported (1-7) in patients with a palliative systemic to pulmonary artery shunt and in patients with a prosthetic valve.

Alpha streptococcus (viridans) and coagulase-positive Staphylococcus aureus continue to constitute the two most frequent etiologic agents of infective endocarditis. Inappropriate administration of antibiotics to a febrile adult with congenital heart disease is often responsible for culture-negative endocarditis or for partially treated patients. Culture-negative endocarditis is a particular concern because of both the delay in diagnosis and the accurate choice of appropriate antibiotics.

Sources of bacteremia. Awareness of potential sources of bacteremia sets the stage for prophylaxis (8). Nevertheless, portals of entry are not apparent in many patients, and when infective endocarditis is due to organisms of low virulence, ≥20% of cases are associated with identifiable medical procedures that cause bacteremia (1). Dental procedures that result in interruption of a surface or gingival bleeding (cleaning, filing or extraction) are common potential portals of entry and are thus most likely to respond to prophylaxis (1-8). Transient bacteremia may accompany nasotracheal intubation or suctioning, surgery involving respiratory mucosa and introduction of a rigid bronchoscope. Fiber-optic bronchoscopy has minimal risk of bacteremia unless there is an untreated airway infection (8). Low level bacteremia occurs during upper gastrointestinal endoscopy or transesophageal echocardiography, although the frequency is unclear (8-11). For upper gastrointestinal endoscopy, the American Heart Association (12) does not recommend prophylaxis, and the British Endocarditis Working Party (13) recommends prophylaxis only for patients with a prosthetic valve. A single case of infective endocarditis (streptococcal) was temporally related to transesophageal echocardiography (11). Bacteremia enterococci and gram-negative rods accompanies urologic procedures provided urinary tract infection is present, but bacteremia is infrequent when the urine is sterile (8). Bacteremia seldom accompanies uncomplicated labor and delivery (8), but it is not always possible to predict an uncomplicated delivery. Similarly, a delivery does not qualify as uncomplicated if an episiotomy is required. Postpartum mastitis (especially in nursing mothers) is a source of bacteremia. In the absence of coexisting infection, prophylaxis is not recommended for urethral catheterization, dilation and curettage, uncomplicated vaginal delivery, Caesarean section, therapeutic abortion, sterilization procedures and insertion or removal of intrauterine devices (13). The incidence of infection after insertion of an intrauterine device is about 2.5% annually; these devices should be avoided in women at risk of infective endocarditis (8).

The American Heart Association (12) takes the position that "prophylactic antibiotics are not required in diagnostic cardiac catheterization and angiography because with adequate aseptic techniques, the occurrence of endocarditis is extremely low," but a 3% incidence of infective endocarditis was reported (6) in >300 pediatric patients undergoing cardiac catheterization. The risk of infective endocarditis accompanying cardiac surgery is related less to the underlying heart disease and surgical technique than to perioperative predisposing factors for bacteremia, such as endotracheal intubation and urethral catheterization.
Prevention of infective endocarditis. Good daily oral hygiene is emphasized. Foods that induce caries should be minimized or avoided. The teeth should be cleansed by twice daily brushing with a soft bristle toothbrush and regular gum stimulation and flossing. A dentist or hygienist should be seen twice yearly. It is believed that formulation of drinking water plays a favorable role. The daily "gingival degemizing" with certain mouthwashes or hydrogen peroxide may decrease plaque formation and gingivitis. Meticulous skin care is also to be emphasized. Patients should meticulously avoid squeezing pimples or pustules and should desist from nail biting or picking with the accompanying risk of paronychia and periungual infection. Prophylactic advice best begins in childhood to instill good habits.

Chemoprophylaxis requires direct medical intervention and advice. To be successful, recommendations must be not only bacteriologically effective, but also acceptable to the patient and physician in terms of cost, ease of administration (route and duration) and few if any side effects. Most patients in whom oral penicillin was previously recommended, that choice remains acceptable. However, risk categories have been rendered less important with the current recommendation of oral amoxicillin. Details regarding the preceding recommendations are available in the current American Heart Association statement (12) on prevention of bacterial endocarditis and antiinfective endocarditis prophylaxis.

References

Assessment of Genetic Risk in Congenital Heart Disease

EDMOND A. MURPHY, MD, REED E. PYERITZ, MD, PhD

Most studies on the genetics of congenital heart diseases have devoted much effort to traditional questions, questions that are often not central to the issue. There has been little attention to defining and exploring the fundamental problems to which coherent inquiry can be directed. After long inaction, interest in the genetics of congenital heart disease is slowly reviving. The approaches are on several quite different fronts: cytogenetic, biochemical, analytic, statistical, mathematical and clinical. Though quite diverse and not easily unified, they are by no means irreconcilable. The prospects are promising, exciting, even revolutionary; but at this stage, the knowledge is germinal only and not ready for clinical application. We (1) recently reviewed some of this knowledge.