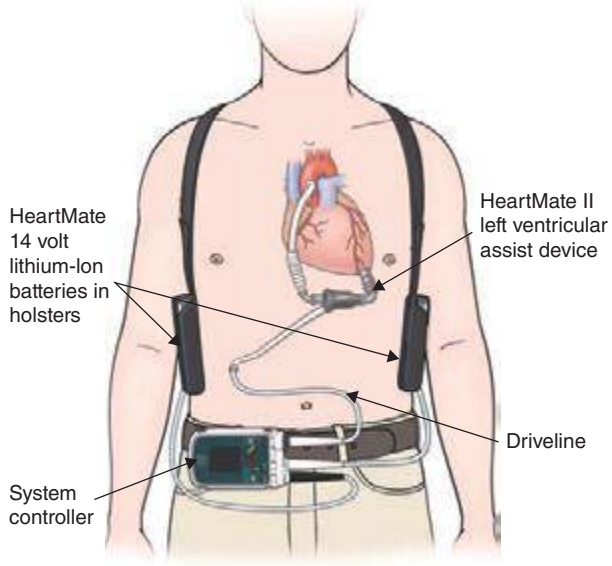

FIGURE 281-2 Diagram of HeartMate II left ventricular assist device (LVAD). *(Reprinted with permission from Thoratec Corp., Pleasanton, CA.)*

The HeartWare Ventricular Assist System with the HVAD pump (HeartWare Inc., Framingham, MA) is the first third-generation device to be granted FDA approval for use in patients as a bridge to transplantation. The device is a centrifugal pump that is housed completely within the patient's pericardial cavity and provides adequate support for many patients.

RESULTS

The use of these devices in the United States is limited mainly to patients with post–cardiac surgery shock and to those who are bridged to transplantation. The results of bridging to transplantation with the available devices are quite good, with nearly 75% of younger patients receiving a transplant by 1 year and having excellent posttransplantation survival rates.

Publication of the REMATCH (Randomized Evaluation of Mechanical Assistance in the Treatment of Heart Failure) trial in 2001 documented a somewhat improved survival rate in patients who had end-stage heart disease, were not candidates for transplantation, and were randomized to a pulsatile LVAD (albeit with a high rate of complications, especially neurologic issues) as opposed to continued medical therapy. This result led to renewed interest in use of the devices for nonbiologic permanent replacement of heart function as well. Subsequently, this device was supplanted by the HeartMate II axial-flow device, which has dramatically improved the survival of patients with severe end-stage heart disease in whom medical therapy has failed. The patients who had this device implanted had a 2-year survival rate of 58%, whereas the survival rate for patients in the medically treated arm of the original REMATCH trial was only 8%. More recent experience has shown that the mean survival period of patients with a continuous-flow LVAD for destination therapy is approaching 5 years.

Several studies have evaluated the benefit of LVAD therapy as a bridge to transplantation. The most recent data come from a series of 140 patients who underwent implantation of a HeartWare HVAD. Of these patients, 94% achieved the principal outcome (defined as survival to transplantation, recovery of heart function, or ongoing device support) at 180 days. With increased experience and improved outcomes using LVADs as a bridge to transplantation, the ability to maintain end-organ function and limit the progression of pulmonary hypertension—or even to decrease pulmonary vascular resistance—makes mechanical unloading a more attractive option than continued inotropic support. The early bridge-to-transplantation experience demonstrated reduced posttransplantation survival compared with medical management; however, more recent experience has shown equivalent outcomes following transplantation. This result is likely secondary to a trend toward earlier device implantation—i.e., prior to the onset of irreversible end-organ damage.

282 Congenital Heart Disease in the Adult

Jamil A. Aboulhosn, John S. Child

Over a hundred years ago, Sir William Osler, in his classic textbook *The Principles and Practice of Medicine* (New York, Appleton & Co, 1892, pp 659–663), devoted only five pages to "Congenital Affections of the Heart," with the first sentence declaring that "[t]hese [disorders] have only limited clinical interest, as in a large proportion of cases the anomaly is not compatible with life, and in others nothing can be done to remedy the defect or even to relieve symptoms." Fortunately, in the intervening century, considerable progress has been made in understanding the basis for these disorders and their effective treatment.

The most common birth defects are cardiovascular in origin. These malformations are due to complex multifactorial genetic and environmental causes. Recognized chromosomal aberrations and mutations of single genes account for <10% of all cardiac malformations. Congenital heart disease (CHD) complicates ~1% of all live births in the general population—about 40,000 births/year—but occurs more frequently in the offspring (about 4–10%, depending on maternal CHD type) of women with CHD. Owing to the remarkable surgical advances over the last 60 years, >90% of afflicted neonates and children now reach adulthood; women with CHD may now frequently successfully bear children after competent repairs. As such, the population with CHD is steadily increasing. Women with CHD are at increased risk for peri- and postpartum complications, but maternal CHD is generally not considered an absolute contraindication to pregnancy unless the mother has certain high-risk features (e.g., cyanosis, pulmonary hypertension, decompensated heart failure, arrhythmias, aortic aneurysm, among others). Consultation with an adult CHD expert is warranted for all females with CHD who desire to become pregnant.

Nearly one and a half million adults with operated or unoperated CHD live in the United States today; there are now more adults than children with CHD in the United States. Because true surgical cures are rare, and all repairs—be they palliative or corrective—may leave residua, sequelae, or complications, most require some degree of lifetime expert surveillance. The anatomic and physiologic changes in the heart and circulation due to any specific CHD lesion are not static but, rather, progress from prenatal life to adulthood. Malformations that are benign or escape detection in childhood may become clinically significant in the adult. Unfortunately, the growing number of adults with CHD has not been paralleled by an adequate increase in the number of specialists and