INTRODUCTION

Mankind has long recognized the heart as vital to sustaining life—often romanticizing it as the repository of the soul and the seat of the emotions—but we did not have the ability to repair it surgically until a relatively short time ago. Open-heart surgery seems so commonplace now that it is sometimes difficult to remember that it was not widely available until the mid-1970s. Curiosity and experimentation, however, have existed for centuries.

Although the first successful operation on the living heart was not until the middle of this century, the recorded history of open-heart surgery goes back as far as about 400 B.C., when Greek physicians provided an account of the workings of the aortic and pulmonary valves. A second-century-A.D. Greek physician, Galen of Pergamon, who was an active dissector of human cadavers, described the heart in detail, but with some notable inaccuracies that were not cleared up until the writings of Andreas Vesalius in 1534.

It was not until the pioneering work of William Harvey, the 17th-century English physician, that blood circulation and the role of veins and arteries were understood. Prior to Harvey's famous dissertation, De Mortu Cordis, it was generally thought that the blood ebbed and flowed like whimsical tides, controlled by the consumption of food.

The first recorded successful heart surgery performed on a living human being was in 1896, when a Frankfurt physician sutured a wound in the heart of a young German soldier. Great strides have been made in this field of surgery since the removal of shell fragments from the hearts of American soldiers in World War II and the first repairs of inborn (congenital) abnormalities in 1945.

Surgical technique in the early 1900s was far more advanced than the ability to keep patients alive. Ability to operate, however, was limited by the inability to operate on a heart that was still beating.

The difficulty of operating on a beating heart was not resolved until the mid-1950s and early 1960s. In early experiments, scientists found they could stop and restart the heart, but this left less than three minutes in which to operate before irreparable brain damage occurred. Philadelphia's John Gibbon was one of the doctors working on a solution: a machine that would take over the circulation of the blood. His first model was tested in animal experiments in 1931, but it was not until 1953 that Gibbon was able to perform a successful operation on a human patient using total cardiopulmonary bypass.

Taking over the blood circulation involves far more than simply pumping blood. The machine has to resupply the oxygen that the body's cells have removed from the red blood cells and pump the blood at sufficient pressure to supply all the organs in the body, without damaging the white or red blood cells or the platelets carried by the circulating blood.

It was not until the mid-1970s that the machines became sufficiently sophisticated to achieve safe, widespread use. Today's bypass machines can maintain the patient's circulation for many hours without serious side effects. Nevertheless, cardiothoracic sur-
METHODS OF TREATMENT

Oxygenator

Figure 25.1

The heart-lung machine takes blood from the patient’s heart to a reservoir from which it travels through a series of thin-walled membranes. These membranes allow oxygen to enter the red blood cells. After impurities are filtered from the blood, it is pumped to the aorta for distribution to the body.

geons seek to keep operating time to a minimum in order to reduce even the small chance of ill effects. The cardiopulmonary bypass machine (popularly known as the “heart-lung machine”) works as follows (see Figure 25.1). Blood is passed along a tube from the patient’s heart (usually from the right atrium) to a pump that pushes blood through a series of thin-walled membranes that duplicate the lung’s method of allowing oxygen to enter red blood cells. The oxygenated blood is passed through a series of fine-meshed filters to trap any impurities. The blood is then redirected to the aorta, the body’s largest artery, where it is distributed to the arteries around the body.

The other technical innovation that allowed doctors to operate on the heart for an extended period of time was the introduction of safe preservation techniques, using extremely cold temperatures (hypothermia), for a heart that has been stopped. In the late 1950s, Dr. Norman Shumway showed that the heart’s demand for oxygen could be considerably reduced and the heart muscle cells preserved if the heart was immersed in a cold salt (saline) solution.

During today’s heart operations, three methods of hypothermia are combined: cooling of the entire body by cooling the blood in the heart-lung machine, immersing the heart in cold saline, and injecting a cold solution of potassium directly into the heart. The high concentration of potassium instantly stops the heart’s electrical activity. By slowing down the heart muscle’s demand for oxygen, the surgeon is able to preserve heart muscle (myocardial) cells for as long as six hours. The enhanced techniques of cell preservation have also made it possible to preserve a donor heart for cardiac transplantation during long-distance transport.

OPEN-HEART SURGERY

A description of a coronary artery bypass operation is somewhat representative of other types of heart surgery that follow similar patterns, although there is obviously some variation, depending on the particular surgery. A primary difference between coronary artery bypass and most of the other surgeries is that the heart chambers are not opened in bypass surgery—as they would be for valve replacement, for example. Surgery in which the heart’s chambers are entered always carries some additional risk; this is primarily due to the increased possibility of air entering the heart, and surgeons take extra care to avoid this.

Once a physician has determined that surgery is necessary, the preparation begins. (See box, “Before and After Open Heart Surgery What to Expect for Adults.”) On the morning of surgery, the patient is given a mild tranquilizer to reduce any anxiety related to the operation. Electrodes connected to an ECG monitor are then attached to the patient’s back to allow constant monitoring of the heart’s electrical activity during the operation. Following the administration of a local anesthetic, intravenous (IV) lines are inserted into the veins of the arm or wrist. These IV lines allow the anesthesia team to administer anesthetics directly into the bloodstream and to replenish body fluid with salt solution. One of these lines is threaded up the vein all the way to the vena cava (a large vein near the heart) to allow administration of medication directly to the heart. Another IV line allows measurement of the pressure and oxygen level in the arteries.

A special balloon-tipped catheter, known as the Swan-Ganz catheter, is inserted into a neck vein and threaded down into the cavity of the right heart and through the right ventricle; blood flow carries it
Before and After Open Heart Surgery: What to Expect for Adults

Before having open heart surgery, the patient will meet with the cardiothoracic surgeon, who will already have reviewed the tests (such as films from the angiography) and will explain the results of these tests, what the surgery entails, the benefits the patient can expect from surgery, and, especially, the risks of surgery. This meeting provides the patient a chance to voice fears, ask questions, and perhaps look over the angiogram film to develop a better understanding of why the operation is necessary. The hospital where the surgery is to take place will most likely provide easy-to-follow literature on the particular surgery and informative meetings for the whole family to discuss pre- and postoperative care.

Approximately two weeks before the surgery, the patient may be instructed to stop taking any medications that might affect the ability of the blood to clot, including aspirin, dipyridamole (Persantine), and warfarin (Coumadin). The patient should continue taking beta blockers and calcium antagonists if he or she is on them and can continue use of nitrates to relieve chest pain. The day before the operation, the patient will be expected to check into the hospital and undergo a battery of tests—chest X-ray, ECGs, blood tests, urinalysis—even though he or she may recently have had all these tests. This is an important step to make sure that there has been no change in the status of the patient’s health and to ensure that no small but important detail has been omitted. The patient will be asked about medications and alcohol, cigarette, and recreational drug use. It is vital that the answers be forthright and complete, as any of these substances can adversely affect the healing process.

The night before the surgery, the patient will be asked to shower in order to reduce the bacteria on the skin. No food will be allowed after midnight, because anesthesia is safer on an empty stomach. The anesthesiologist will usually visit the patient the night before in order to explain what the anesthesia does and how the anesthesia team takes care of the patient’s breathing with the respirator. The chest area will be scrubbed clean and washed with an antiseptic solution, and body hair will be shaved where necessary.

On the morning of the surgery, the person will be given a mild tranquilizer to reduce anxiety about the operation. Monitoring electrodes will be attached, and local anesthetic will be administered before placement of intravenous lines. Catheters (thin tubes) that perform such duties as collecting urine and monitoring blood pressure are inserted. The first few seconds of the administration of the general anesthetic should be the last part of the operation the patient remembers.

After the surgery, the patient is taken to the intensive care unit (ICU), where specialized nursing care is available around the clock and sophisticated instruments monitor the heart’s electrical activity, blood pressure, temperature, and other vital signs. The patient’s family will generally be allowed to visit at this time, although the patient will be groggy from the anesthetic and unable to speak because of a tube in the windpipe (endotracheal) that helps the person breathe with a respirator. The ICU experience can be disorienting, as there is little to differentiate day and night, and the patient will drift in and out of consciousness. The nurses are specially trained in communicating by touch and signboard, and they will do everything possible to make the patient comfortable.

Each person recovers at his or her own speed, and much depends on the nature of the surgery. In most cases, the patient will be taken off the respirator the morning after surgery, and the catheters and intravenous lines will be removed within two days. The patient can then hasten his or her recovery by following the guide lines for inflating the lungs (by sucking on a special lung-exercising device) and by making an effort to become mobile, especially by walking. (See Chapter 28 for more information on cardiac rehabilitation.)

through the pulmonic valve into the lung or pulmonary artery. The Swan-Ganz catheter provides an accurate reading of the pulmonary arterial blood pressure (based on the pressure at the tip of the balloon) and indicates how well the heart is functioning. A sensitive temperature probe at the tip of this catheter can also tell the surgical team how well blood is circulating.

A Foley catheter is inserted into the patient’s bladder before the operation. The amount of urine collected by this catheter is a sign of how well the patient’s kidneys are functioning and whether the kidneys are receiving sufficient oxygenated blood.

Anesthetic agents are then administered directly into the veins. These agents have three functions: to block pain and induce drowsiness, to relax the muscles and prevent the person from moving and jerking during the operation, and to cause temporary amnesia so the person is not disturbed by a detailed recollection of the operation. The anesthesiologist
carefully monitors the patient’s vital signs throughout the operation, adjusting the dosage of medications and anesthetics appropriately.

Once the patient has been anesthetized, a tube (endotracheal) is inserted into the patient’s windpipe. The tube connects to the respirator—a bellows-like instrument that performs the work of breathing for the patient. Another tube (nasogastric, or NG) is also inserted to collect stomach fluids that might otherwise nauseate the anesthetized patient.

An anticoagulant, heparin, is also administered at the start of the operation. This “blood-thinner” prevents clots, or emboli, from forming and helps to protect the patient from a stroke. The effects of the heparin will be reversed at the end of the operation by the administration of another drug, protamine, that encourages coagulation.

Once the patient has been prepared, the surgeons begin. For open-heart surgery, the chest is cut open at the midline of the breastbone (sternum) and the breastbone is separated. The chest is then gradually pried open with special retractors to reveal the lungs and, between them, the tough sac of tissue (the pericardium) that protects the heart.

If the internal mammary artery (an artery that supplies blood to the chest wall) is to be used for bypass grafting, at this time it will be gently separated from the chest wall. During the opening of the chest, another surgeon will have been working on the patient’s legs to remove several usable lengths of a vein (approximately 20 centimeters, or 8 inches, for each bypass). These lengths of vein are about the diameter of a drinking straw. Later in the operation, the surgeon will take each in turn and sew one end into a tiny hole punched into the aorta and attach the other end to a coronary artery, thereby providing the bypass pathway around each narrowing in the coronary arteries. (See Figure 25.2.)

Once the sac covering the heart has been opened, the surgical team sets up the heart-lung machine. Several plastic tubes are hooked up to the machine. When it is clear that the heart-lung machine is providing adequate circulation, the aorta is clamped, and the heart is stopped with an injection of cold potassium solution directly into the aorta. The outside of the heart is also bathed in a cold salt solution to further induce hypothermia. The patient is now on total bypass; his or her blood circulation has been completely taken over by machine. The surgeons now can attach the bypassing vessels. (Or, if this is an operation other than a coronary bypass, the chambers of the heart can be opened and the appropriate surgery performed.)

Once the direct surgery on the heart has been performed, the aortic clamp is removed, and the blood is gradually warmed. The heart may begin beating by itself, or the surgeon uses a brief shock to restore electric activity. Pacemaking wires are placed to allow electrical control of the heart rate. When the heart supports its own blood circulation again, the patient can be taken off the heart-lung machine, any bleeding can be stopped, and incisions can be closed. After the patient is taken off the bypass machine, protamine is injected to reverse the effects of the heparin by restoring the normal clotting ability of blood, and the patient is transferred to an intensive care unit.

**Figure 25.2**
*This illustrates the two main types of coronary bypass grafts: saphenous vein and internal mammary artery. On the left, a section of saphenous vein from the leg is sutured between the aorta and a coronary artery, bypassing the blockage. The left internal mammary artery, normally found in the chest, is redirected to bypass another blockage.*

**CORONARY ARTERY BYPASS SURGERY**

In the last few years, coronary artery bypass grafting has become not only the most common heart operation, but also one of the most frequently performed surgical procedures. In 1988, 320,000 bypass operations took place in the United States. The procedure has become so entrenched that it is easy to forget the first such operation was performed as recently as
1967, at the Cleveland Clinic, when Dr. Rene Favaloro used a vein from a leg to bypass a blocked coronary artery. The basic operation has remained much the same, but improvements in surgical technique, in heart preservation during heart-lung machine use, and in the understanding of when and how to also use an artery from the chest wall as a graft have led to longer-lasting grafts, reduced death rates, and increased ability to provide relief for older and sicker patients.

This surgery is usually elective (except for the emergencies that may occur during a threatened heart attack), and the patient often plays a large role in deciding both when and whether to have the operation. By the time most patients are considering the operation, they will have experienced symptoms of heart disease and already have made life-style changes and been treated with medication.

It is useful to distinguish between surgery that is required based on the location of blockage in an artery (anatomic indications) and surgery that is done to improve the heart’s function and relieve symptoms (functional or symptomatic indications). Anatomic indications are determined by cardiac catheterization and other tests. Currently, coronary angioplasty may provide an effective alternative to surgery in certain instances. (See Chapter 24.)

Several types of artery narrowing call for surgery. The left main branch of the coronary arteries is a short section leading from the aorta (like the main trunk of a tree) that divides into the circumflex and left anterior descending arteries. If the left main branch is narrowed or constricted (stenotic), it is of particular concern, because the blood supply to much of the heart could be suddenly reduced. People with untreated left main artery disease have an approximate death rate of 50 percent over a five-year period.

The other major artery, which also divides into smaller branches, is the right coronary artery. It supplies blood mainly to the right side of the heart and the underside of the left ventricle. When this artery becomes narrowed or blocked, it is usually not as serious as when the left arteries are affected; surgery is usually not necessary if right artery blockage is the only major problem.

Triple-vessel disease refers to significant (greater than 70 percent) narrowing of the interior of all three coronary arteries. Without coronary bypass, patients with triple-vessel disease have a relatively poor prognosis, particularly if heart function is reduced.

Patients who have suffered damage to the muscle of the main pumping chamber from a heart attack may also be considered candidates for coronary artery bypass surgery. This chamber, the left ventricle, plays a crucial role in pumping arterial blood to the rest of the body (including the coronary arteries themselves), so its efficiency must be maintained. Left ventricle efficiency is usually determined by the amount of blood squeezed out with each beat (the ejection fraction). Coronary artery bypass surgery is advisable when this becomes reduced to a less than adequate level.

Choosing surgery to improve function or relieve symptoms is a more subjective decision; the person’s own feelings about life-style restriction may be an important criterion. When chest pain (angina) occurs with unusual frequency or at rest, despite continuing use of medication, surgery may be strongly urged: These symptoms are often warning signs of an impending heart attack. However, some patients with less serious angina that may not actually be getting worse may also opt for surgery, as they may be intolerant of the medications or of the restrictions imposed upon their work and leisure activities.

The principle of coronary artery bypass surgery is to provide a new blood supply for sections of the heart muscle whose own supply of arterial blood is restricted by a blocked artery. The conduit that supplies the new route for blood can be a section of a vein that has been removed from the leg (saphenous vein) and attached to the aorta and the coronary artery to bypass the narrowed section. Another possible conduit source is an artery called the internal mammary artery, a blood vessel that usually supplies blood to the chest wall. There is strong evidence that a bypass using a section of this artery is less susceptible to becoming blocked in the future. Only 60 percent of grafts using a vein are still open after ten years as opposed to more than 90 percent of grafts using an artery.

Surgery using internal mammary artery grafts takes slightly longer because the detaching and reattaching process is more complicated, so there was some early resistance to their use. Nearly all bypass surgery patients who have multiple grafts now have at least one using the internal mammary artery. There are, however, some reasons to avoid using it; these will be considered by the surgeon.

It should be noted that internal mammary artery grafts are often performed in conjunction with saphenous vein grafts. Surgeons believe that providing more new conduits to replenish the blood supply increases the chances of a successful long-term outcome. This is the reason for triple, quadruple, and even quintuple bypasses (referring to the number of new conduits created). The number of bypass grafts...
METHODS OF TREATMENT
does not necessarily indicate the severity of the disease. Too often, patients are led to believe that a quadruple or triple bypass implies that they may have a terrible condition. This is not true. Several of the blockages bypassed may have been relatively “minor” ones.

The patient who has received a coronary artery bypass graft can expect considerable relief from symptoms, and, in many cases, increased lifespan. It should be remembered, however, that the graft vessels are subject to fatty blockage at an increased rate, so care must still be taken to reduce the risk factors that caused the original blockage. (See Chapter 3.) The surgery sets the disease back to an earlier stage; it does not “cure” atherosclerosis. A certain number of patients—especially those whose original bypass surgery occurred more than ten years ago—will find that they are candidates for a second bypass (or “re-op”). This second bypass operation carries only slightly more risk than the first and is an important option for people whose grafts have become constricted and whose symptoms have recurred.

Newer and better surgical techniques now allow surgeons to operate safely and effectively on some people who only a few years ago might have been considered too old or too sick for surgery.

VALVE REPLACEMENT

The heart’s valves perform the vital function of maintaining blood flow in the correct direction. The mitral valve directs the flow of blood from the left atrium into the left ventricle, and the aortic valve allows blood to pass from the left ventricle into the aorta. The tricuspid and pulmonary valves perform the equivalent tasks on the right side, but are under considerably less pressure than the valves on the left side and—although they may suffer from similar disorders—are less likely to be so severely impaired as to require surgery. (See Chapter 13.)

Problems with the heart valves and their functioning can be of two kinds: narrowing (stenosis), when the valve opening is constricted and blood flow reduced, and regurgitation, when some of the blood leaks back into sections of the heart from which it has just been expelled because the valve leaflets do not close properly. A poorly functioning valve in which the leaflets neither open nor close properly may cause both problems.

While poorly functioning valves most often can be detected by listening with a stethoscope, a definitive diagnosis of valve disease usually requires such tests as X-rays and an echocardiogram. The decision to operate and correct a valve abnormality will depend on whether the condition is life-threatening and to what extent it has affected the person’s life-style. Most problems with valve disease are not of an urgent nature, and the decision does not have to be rushed.

The exception is when the valve opening between the aorta and the left ventricle is blocked (aortic stenosis), which surgeons consider an urgent problem if symptoms occur (such as chest pain, faintness, and shortness of breath) and if the narrowing is severe. About 50 percent of such patients die within four years without surgery. When the aortic valve does not close properly (aortic insufficiency), the decision to operate depends on the degree of damage that shows upon various tests and on whether the person has symptoms. (Less active people may be willing to curb their activity—thus reducing demand on the heart—in order to keep their symptoms to a minimum and avoid surgery.) Poorly functioning aortic valves can result in enlargement of the left ventricle, a condition that must be monitored carefully.

When the valve opening between the upper and lower chambers of the left side of the heart (mitral valve) becomes severely blocked (mitral stenosis), it usually requires surgery. Because the narrowing of the valve opening may cause blood to back up into the lungs, careful monitoring of symptoms such as shortness of breath is required, and surgery maybe called for to prevent serious heart failure. When the mitral valve closes improperly (mitral insufficiency), the desirability of surgery is usually determined by how severely the symptoms affect the patient’s lifestyle and how well they can be controlled by medical treatment.

Heart valve disease can be surgically treated in three ways: Constricted openings can be enlarged with a balloon catheter; the injured valve can be surgically reconstructed; or the valve can be replaced, either with an artificial valve or with a healthy valve from a pig’s heart.

A balloon catheter can be used to alleviate mitral stenosis in some cases. (See Chapter 24.) Mitral valve problems associated with rheumatic fever usually occur because the leaflets stick together. Sometimes simply opening the leaflets enough to separate them from each other is all that is required to solve the problem. To accomplish this, the balloon catheter is threaded through the valve. When it is in place, the balloon at the tip of the catheter is inflated gently until
it enlarges the opening. This procedure is rarely an option for aortic stenosis, except in people whose life expectancy is limited because of other diseases. Balloon dilation is not effective, and there is a risk that the calcified leaflets will break off and enter the bloodstream, causing a stroke.

Surgical repair can be performed on the mitral valve, particularly to relieve mitral insufficiency. The procedure followed is the same as for other forms of open-heart surgery. The heart is stilled and the left atrial chamber opened, then the leaflets are reconstructed to allow the valve to close properly. If the leaflets of the mitral valve have become stuck together as the result of rheumatic fever, the surgeon can separate them and suture any damaged edges to ensure that they close efficiently. The advantage of this surgery is that there is no new valve to wear out, nor does the patient ordinarily have to follow a regimen of blood thinners as he or she would with a mechanical heart valve replacement.

Artificial valves have been in use since 1952, when Charles Hufnagel successfully replaced a patient’s aortic valve with a caged-ball artificial valve. Some mechanical valves are of the “tilting disk” variety, while others are of the “ball-and-cage” variety. (Figures 25.3A–C show some commonly used artificial valves.) The latter consist of a metal ring covered in Dacron and a thin metal cage. Inside the cage is a ball, which exactly fits the dimension of the ring. Blood flowing in the correct direction moves the ball away from the ring towards the cage and flows by uninterrupted. When the blood begins to flow in the opposite direction, it pushes the ball back into the ring, creating a tight seal that prevents leaking. The tilting disk valves work somewhat along the principle of a hinged door that is opened due to pressure from one side and shut tightly due to pressure from the opposite side. Mechanical valves make a clicking noise with each heartbeat, but the patient usually can not hear this.

Valve replacement is performed during open-heart surgery. Artificial valves are carefully sutured or sewn into the ring surrounding the valve opening, completely replacing the natural valve. (See Figure 25.3D.) Approximately 80 percent of patients who survive the first postoperative year are able to return to normal activity, even though they may previously have been severely restricted by breathlessness from heart failure or by fainting spells and angina. The downside of the operation is that it carries with it a 5 percent mortality risk (somewhat higher than for coronary artery bypass surgery), partly because of the possibility of a stroke caused by loosened calcium
METHODS OF TREATMENT

Valves taken from pig hearts do not ordinarily require the patient to take anticoagulant medication for more than six to eight weeks following surgery. The disadvantage of these replacement valves is that they do wear out and may have to be replaced about ten years. They wear out gradually, so there is little risk of a sudden episode of heart failure. It should be noted that valve tissue does not cause rejection problems as seen with heart transplantation. Recent studies have shown that the natural valve compares favorably with the artificial valve over a ten-year period; patient longevity may even be enhanced. When comparing the statistics, people should bear in mind not just the length of time a valve will last, but also the problems of infection and anticoagulant drug therapy.

SURGERY FOR HEART RHYTHM DISORDERS (ARRHYTHMIAS)

Arrhythmias are disturbances in the heart’s electrical conduction system, which controls both the sequence and the frequency of heartbeats. These can take the form of harmless “palpitations” or become present as a sign of injury or disease. Irregular or rapid rhythms generally originate in two areas: the ventricles (the lower part of the heart) or the supraventricular areas (the upper part of the heart). Ventricular arrhythmias commonly occur as extra beats and are generally of no importance. In people who have had heart attacks with tissue damage, however, a sustained run of extra beats can be serious. Between the dead muscle and the normal muscle is an unstable “border zone” from which extra beats may arise. If they occur in sequence and at a rapid rate (ventricular tachycardia) or as rapid, irregular, and uncoordinated beats (ventricular fibrillation), urgent treatment is required.

Many people with a history of ventricular tachycardia or fibrillation come to the surgeon after having experienced near “sudden death” and resuscitation. If these arrhythmias have been recurrent and are not controlled by medical therapy (see Chapter 16), further studies may be necessary. Cardiac catheterization and the use of an electric probe may help to locate a problem in the heart muscle. The surgeon then cuts out unstable areas that are causing the arrhythmia. The operation has a mortality rate of around 12 percent, but it is effective in preventing these serious
Arrhythmias can also occur in people who are born with extra conducting pathways. The impulse from the atria short-circuits the normal pathway and activates the ventricle sooner than normal. The short-circuiting is detectable on an electrocardiogram. The most common type of abnormal pathway result is Wolff-Parkinson-White (WPW) syndrome, in which an additional conducting pathway bypasses the atrioventricular (AV) node, which separates the atria from the ventricles. The AV node normally acts as a regulating block for electrical impulses passing from the atrium to the ventricle, and the additional pathway can allow abnormally frequent electrical signals from the atria to setup arrhythmias in the ventricles.

Patients with Wolff-Parkinson-White syndrome are, for the most part, young and have otherwise normal hearts. Most of them have few symptoms, but may experience episodes of rapid heartbeats, some of which may be severe. The syndrome was formerly treated with medication, which was not completely effective in preventing recurring arrhythmias. Today, if symptoms are recurrent and severe, the extra conducting pathway can be removed during open-heart surgery. This operation provides a cure, and the mortality rate during surgery is low.

**REPAIR OF ANEURYSMS**

An aneurysm is a severe weakening in the wall of an artery or organ. The weakened area balloons out. In addition, the bulging area may either press on and disrupt the function of other vital organs or burst and hemorrhage.

There are two kinds of aneurysms that may directly affect the heart: left ventricular aneurysm and aortic aneurysm. Left ventricular aneurysms may appear following a heart attack (myocardial infarction, or MI). This is a relatively uncommon occurrence. It is a bad sign, because it indicates that a lack of oxygen has caused extensive damage to the muscular wall of the left ventricle. Surgeons operate on left ventricular aneurysms if they are accompanied by arrhythmias, congestive heart failure, or blood clots. The operation is performed through the area of the heart attack, and the surgeon seeks to remove or repair the weakened area of the ventricle wall. This operation allows the ventricle to function more effectively (although heart muscle that has been destroyed cannot be restored) and can usually give the person a reasonable life expectancy.

An aneurysm in the aorta may be surgically repaired by replacing that section with a prosthetic graft, usually made of Dacron. This surgery can also have decent results, but it carries a mortality risk of approximately 10 percent.

**HEART TRANSPLANTS**

The idea of replacing a diseased heart with a healthy one has long fascinated surgeons. The earliest heart transplant was carried out in 1960 at the University of Chicago. Doctors successfully implanted a heart into a dog’s neck; a beat was sustained for several hours. The first demonstration of the possibility that a transplanted heart could completely take over the circulation was made by a Russian physician in the 1950s.

Transplants in which the transplanted heart is placed in the normal anatomical position and the diseased heart removed were soon recognized as being desirable. The first such transplants were done in the early 1960s using animals. In 1964, a dying 67-year-old man’s defective heart was replaced with that of a chimpanzee; a beat was sustained for a few hours. Three years later, Christian Barnard became a worldwide celebrity as the first surgeon to successfully transplant a heart from one human being to another.

These initial attempts were dampened by poor survival rates for the patients. Several more advances were required before heart transplantation became a truly viable method of treatment in the early 1970s.

**PATIENT SELECTION**

Careful selection of transplant recipients has led to an improvement in the success of these operations and in the more effective use of the limited pool of available donor organs. A recipient of a heart transplant should be someone who is less than 60 years old (there is no lower age limit), who is suffering from end-stage congestive heart failure (see Chapter 14) with severe symptoms, and for whom survival with-
out the transplant is extremely limited. The main causes of this type of severe heart failure are coronary artery disease (see Chapter 11) and heart muscle disease (see Chapter 15), although some transplants are now being done for congenital heart disease (see Chapter 20).

A transplant is usually not advised when another major systemic disease exists, such as a malignancy, lung disease, collagen vascular disease (such as lupus), or insulin-requiring diabetes mellitus.

Rigorous criteria for donor selection have also greatly increased the success of heart transplantation. The suitable donor has to be less than 35 years of age and to have sustained brain death (usually from cerebral trauma, for example a head injury or stroke) with no evidence of chest injury. Signs of infection, a history of cardiac illness, or prolonged use of cardiopulmonary resuscitation would rule out the donor’s organ.

When a donor heart that meets these criteria becomes available, the next step is to match it promptly with an appropriate recipient. Matching is now done through regional organ banks, and the donor organ is flown to the hospital where the recipient is concurrently being prepared to receive it. Matching is done on the basis of blood type, and there is a general requirement that the donor be approximately the same size or larger than the recipient. The reason for the latter requirement is to avoid exposing the donated heart to the increased blood pressure requirement that is present in a larger person.

THE OPERATION

When an appropriate donor/recipient match has been made, based on careful screening techniques, the donor’s chest is opened. The heart is carefully examined for previously unrecognized injury, is cooled, and then is removed from the chest. It is particularly important that the correct protocol for the donor operation be followed carefully.

The recipient is also carefully prepared for surgery; the surgeons try to time the use of the heart-lung machine with the arrival of the donor organ. The recipient’s heart is not removed in its entirety—some portions of the atria, or thin upper chambers, are preserved. (See Figure 25.4A.)

Transplanting only part of the heart greatly simplifies the operation. The main sections that need replacing are the ventricles (whose muscular walls do most of the heart’s work), the valves, and only part of the atria. (See Figure 25.4B.)
Care is taken throughout the operation to keep the donor heart cool and to keep air from being trapped in the heart chambers. (See Figure 25.4C.) When the connections are complete (see Figures 25.4D–F), the patient’s new heart is gradually warmed and resuscitated, then taken off the bypass machine. As in most other forms of heart surgery, atrial and ventricular pacing wires are attached to the outside of the heart to allow for rapid treatment of any rhythm disturbances after the chest has been closed.

Two major issues are of concern after surgery preventing and diagnosing the possible rejection of the donor heart by the recipient’s body, and avoiding infections. The high mortality and failure rate of heart transplants in the late 1960s arose largely from a lack of knowledge in properly managing these potential difficulties.

Graft rejection results when the body’s immune system recognizes the donated organ as an alien substance and begins attacking and trying to destroy the “intruder.” Because perfect matching of donated tissue with the recipient’s tissue is simply not feasible for these kinds of operations, the immune system needs to be suppressed. The discovery of cyclosporine (Sandimmune), a drug that originated in a soil
mold from Norway (dug up by a vacationing researcher), has increased the ability to suppress certain aspects of the immune system that may cause graft rejection, while still allowing the body to maintain its defenses against infection. Cyclosporine, even in small doses, mainly suppresses one kind of immune cell (T-cell lymphocyte), whereas earlier immunosuppressant medications unnecessarily suppressed a broader range. T-cell lymphocytes are important components of the system that tends to surround and get rid of foreign substances. Cyclosporine is generally taken in combination with other immunosuppressant drugs—such as a steroid medication called prednisone (Deltasone) and azathioprine (Imuran)—so smaller doses of each medication can be used.

Improved methods of diagnosing rejection have also led to better long-term management of the body's immune response. Effective testing is essential, because the clinical signs of rejection are generally unreliable: The patient may complain of malaise or have a low-grade fever, but often there are no discernable symptoms. Rejection of the new heart cannot be reliably diagnosed using an electrocardiogram. Cardiac biopsy, performed under local anesthesia by passing a biopsy instrument through a vein and into the heart, is required to allow microscopic study of the tissue. If rejection appears to be taking place, the dosage of cyclosporine can be increased and then lowered again to normal levels once the patient has improved.

Heart transplantation has come to be an effective and beneficial option for some patients who, without the surgery, would have lived for only a few months at best. Approximately 85 percent of patients are still alive after the first year and 65 percent after five years, and complete rehabilitation occurs in about 87 percent of the patients. Patients must be followed very closely after transplantation: Routine clinic visits are necessary with regular biopsies scheduled in order to detect rejection. In addition, cardiac catheterization and coronary angioplasty maybe performed annually to assess the condition of the heart and coronary arteries.

HEART AND LUNG TRANSPLANTS

Transplanting both the heart and one or both lungs is becoming more common for seriously ill patients. Recipients for the double transplant may have sufficiently high blood pressure in their own lungs to cause the failure of a donor heart, or they may be patients suffering from end-stage lung disease with associated heart failure. Distant procurement of lungs has only recently become possible, because the lungs have an extreme sensitivity to oxygen deprivation (ischemia). Lungs can now be maintained for up to six hours outside the body by a new technique of preservation.

The combined heart-lung transplant has a higher mortality than the heart transplant alone, but better preservation of donated organs and improvements in postoperative care have made this an option for selected patients. While technically feasible in many patients, transplantation of a heart or heart and lungs is a major, serious operation with a long period of convalescence. It is a clearly desirable option for only a small number of younger people with advanced, nontreatable heart and lung disease.

PEDiatric HEART surGERY

Approximately 10 percent of the cardiac surgery in this country is performed on infants and children. Problems requiring this surgery are almost exclusively congenital in nature—the children are born with heart defects. (See Table 25.1.) This is in contrast to cardiac surgery performed on adults, which is predominantly for problems acquired during their lifetime, such as coronary artery disease.

Congenital defects vary from straightforward problems, such as a “hole” in the heart, producing mild symptoms or none at all, to complicated malformations that result when a good portion of the heart has failed to develop. Normally, these lead to life-threatening situations. Depending on the heart defect, there are two common conditions that may arise. Children who have more than the normal amount of blood flow to the lungs as a result of a defect may suffer from congestive heart failure. This often causes difficulties in eating, lack of weight gain, rapid breathing, and sweating. A large hole between the left and right main chambers of the heart (ventricles) can produce this.

On the other hand, children with blockage resulting in less blood flow to the lungs may suffer from blueness of the skin (cyanosis). Although they may gain weight and grow well, they are cyanotic because of the low level of oxygen in their blood. These children are often physically limited, and they can suffer from complications such as infections and blackouts.
There are many different congenital heart defects, some of which are quite rare. More common defects account for the majority of clinical problems. It must be remembered that each individual case is unique and may have mitigating factors that require special consideration or treatment.

When there are one or more holes in the wall that separates the right atrium from the left atrium, the child has an atrial septal defect. This results in some shunting of blood from the left side (high-pressure side) of the heart to the right side (low-pressure side) through the hole. This is called a “left-to-right shunt.” There is increased blood flow to the lungs, but pressure in the lungs generally remains low, and the child usually shows no symptoms. The problem is most often diagnosed when a heart murmur is discovered or when there has been an increase in heart size (noted on an electrocardiogram or chest X-ray). While the defect may not produce symptoms in the child, if the hole is large enough it may cause heart enlargement, heart failure, or heart rhythm disorders over the years, significantly decreasing the normal life span.

If surgery is considered necessary, it is usually performed electively when the child is between the ages of 2 and 4. (See box, “Before and After Open Heart Surgery: What to Expect for Children.”) The holes most often can be closed with a suture, but if they are particularly large, a small patch made from the patient’s own pericardial (outside wrapping of the heart) tissue or a Dacron patch can be used. The operation provides a complete cure, and it is extremely rare for the holes to open up again. Antibiotic treatment to prevent infection need not be required for more than a few months after the operation, and the patient should be able to lead a full and active life. A recently developed catheterization procedure that is still considered investigational by the Food and Drug Administration shows promise in treating atrial septal defects. See Chapter 20 for more information.

When there is a hole in the septum, or dividing muscle, between the right and left ventricles, the defect is called ventricular septal defect. As with the atrial septal defect, a considerable amount of blood is pumped back into the low-pressure right ventricle (left-to-right shunting), This condition may exist without symptoms for many years. When they do occur, symptoms can include breathing difficulties and congestive heart failure as a result of the large amounts of blood filling and congesting the lungs.

If the opening is large and the infant is very small and ill, a band maybe placed around the pulmonary artery to restrict blood flow into the lungs and prevent heart failure. Complete repair is then carried out within the first year or two of life, when the band is removed. This course of action is becoming less common; even in very young infants, repair as a first step is considered optimal.

The holes are closed, usually with a small patch of Dacron or Gore-Tex rather than the sutures that are used to close atrial septal defects. The long-term results are very good once the hole is closed, and cure is essentially complete. Postoperative complications are uncommon, but they can include heart rhythm irregularities or the presence of a small residual hole.

The most common form of cyanotic congenital heart disease is tetralogy of Falot. Even so, it is relatively rare. The syndrome includes four abnormalities: a large ventricular septal defect; a displacement of the aorta to the right side so that blood without adequate oxygen enters it from the right ventricle; a
<table>
<thead>
<tr>
<th>Defect</th>
<th>Description</th>
<th>Surgery</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>One or more holes in the wall that separates the right and left atria</td>
<td>Hole(s) closed with a suture or a small patch made from the patient's own tissue or from Dacron.</td>
<td>Operation provides a complete cure; extremely rare for holes to open up again.</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>A hole in the septum between the right and left ventricles</td>
<td>Hole(s) closed with a small patch of Dacron or Gore-Tex.</td>
<td>Cure is essentially complete; rare complications are heart rhythm problems or small residual septal defect.</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>A large ventricular septal defect, a displacement of the aorta so blood with low oxygen enters it from the right ventricle, a thickened right ventricular wall from increased pressure, and partial blockage of the pathway from the heart to the lungs</td>
<td>Ventricular septal defect closed and blockage of blood flow removed from between the right ventricle and the lungs.</td>
<td>Good long-term results; minority experience right-heart failure or heart rhythm disorders requiring additional surgery or continued medication.</td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>Transposition of aorta and pulmonary artery</td>
<td>Most common operation for past few years is arterial switch operation (ASO), in which the aorta and pulmonary artery are reconnected at the proper locations, and the coronary arteries are switched as well.</td>
<td>Short-term results are good; it is too soon to know long-term results.</td>
</tr>
<tr>
<td>Congenital valve disorder</td>
<td>A deformity of any of the heart’s four valves, most commonly aortic valve constriction or narrowing (stenosis)</td>
<td>Valve is opened and repaired and/or replaced with a mechanical valve when child is large enough to accommodate valve of available size.</td>
<td>Results with mechanical valves are good, but anticoagulant medication must be taken indefinitely.</td>
</tr>
<tr>
<td>Complete atrioventricular canal</td>
<td>A central defect connecting all four chambers in the central part of the heart with only one large common atrioventricular valve</td>
<td>Atrial and ventricular septal defect is closed, and two valves are created from the one valve,</td>
<td>Results are generally good; significant number of patients need valve repair or replacement later.</td>
</tr>
<tr>
<td>Defect</td>
<td>Description</td>
<td>Surgery</td>
<td>Prognosis</td>
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<tr>
<td>Tricuspid atresia</td>
<td>A severely underdeveloped right side of the heart (usually with only one adequately sized ventricular chamber)</td>
<td>Fontan procedure diverts all the venous blood return from the body directly into the lungs, bypassing the pumping chambers and returning to the lungs to the one good ventricle to be pumped out,</td>
<td>Does not provide cure but seems to offer long-term relief from cyanosis and heart failure; children require careful lifelong follow-up; long-term consequences remain to be seen.</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>A severely underdeveloped left side of the heart, including the ventricle and ascending aorta</td>
<td>Norwood procedure creates new outflow for heart and shunt to provide blood to lungs until child is old enough to undergo Fontan-type procedure; other option is cardiac transplant.</td>
<td>Results are encouraging in both cases; however, long-term effects of immunosuppressive drugs on growth and development not understood.</td>
</tr>
<tr>
<td>Patent ductus arteriosis</td>
<td>A blood vessel that fails to close after birth and continues to connect the aorta and pulmonary artery</td>
<td>Abnormal connection is closed with small, metal clip; does not require open-heart surgery.</td>
<td>Provides a complete cure.</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>Narrowing of the aorta</td>
<td>Constriction removed through reconstruction of aorta.</td>
<td>Some patients have recurrence of coarctation, but it can be repaired by a balloon dilation procedure in a cardiac cath lab.</td>
</tr>
</tbody>
</table>

thickened right ventricular wall resulting from the increased pressure inside the right ventricular cavity; and partial blockage of the pathway from the heart to the lungs (pulmonary artery).

The symptoms of tetralogy of Fallot are different from those of a simple ventricular septal defect because of the effects of the other complications; blood flow does not increase in the lungs and cause heart failure. Instead, there is a right-to-left shunt of blood—blood without adequate oxygen travels from the right ventricle out the aorta, Not enough blood passes through the pulmonary artery to receive oxygen from the lungs, so children with this defect are often intensely cyanotic; they appear blue from lack of oxygen. Under certain conditions, such as stress, this oxygen deficiency may suddenly become much worse. This results in “cyanotic spells,” which can be serious. Urgent surgery of some kind is necessary.

If an infant has severe symptoms at an early age, placement of a temporary shunt maybe required to provide more blood flow to the lungs. The long-term corrective procedure involves open-heart surgery, including two steps: closing the hole between the two ventricles and opening up the artery from the right ventricle to the lungs. The long-term results of the operation are good; most patients grow normally and have normal lives. A minority of patients have difficulties later in life with heart failure or heart rhythm disorders and may require additional surgery or continued medication.
A very rare congenital defect in which the aorta and pulmonary artery are switched is called transposition of the great arteries. The aorta is connected to the right ventricle instead of the left ventricle, and the pulmonary artery is connected to the left ventricle instead of the right ventricle. So blood that contains a great deal of oxygen that normally should go into the arteries of the body via the aorta is instead pumped back into the lungs. Blood with less oxygen ends up in the liver, brain, kidneys, and so on. These infants are often severely cyanotic.

This transposition can be corrected, but that requires a complicated surgical procedure that sometimes has less than satisfying results. However, newer surgical approaches to correcting this rare abnormality appear promising.

An infant can be born with a deformity of any of the heart's four valves, but the most common congenital valve disorder is aortic constriction or narrowing (stenosis). In this defect, the aortic valve may be small or poorly developed (hypoplastic), or the leaflets may be thickened. The decision on how early to operate depends on the extent of the narrowing and the symptoms. During the operation, the valve will be opened and repaired, if possible; otherwise, it will have to be replaced. There currently are no artificial valves small enough for very young infants, so valve reconstruction has to be done in these cases, with possible replacement at a later stage. Children frequently require placement of mechanical valves, because valves from pig hearts tend to harden rapidly with calcium deposits when placed in someone so young. Mechanical valves require that children take anticoagulant medication indefinitely to keep blood clots from forming in the valves.

A defect seen in children with Down syndrome is a complete atrioventricular canal. In this disorder, instead of two separate atrial-ventricular valves—the mitral on the left and the tricuspid on the right—there is only one large common atrioventricular valve. These children may require several complicated operations to correct this major defect.

In extremely rare cases, infants are born with a severely underdeveloped right side of the heart (usually with only one ventricular chamber instead of two). The defect is referred to as tricuspid atresia. A variety of problems can result, and the infant maybe either cyanotic or experiencing heart failure, depending on the amount of blood flow to the lungs. One or more procedures will be required to relieve symptoms during infancy; the nature of these will depend on the physiology of the individual defect.

Usually at about age 2, the patient can undergo a Fontan procedure, an operation first performed in the early 1970s, which involves diverting all the venous blood return from the body directly into the lungs, thus bypassing the pumping chambers. Blood then returns from the lungs to the one good ventricle, where it is pumped out to the body. Consequently, the one ventricle does the work of two: pumping the blood through the arterial system and then through the lungs in series. Although this operation does not really cure the defect, it appears to offer the best long-term relief available to children with only one heart pumping chamber. The operation does relieve cyanosis and heart failure. These children, however, do require careful lifelong follow-up, and the long-term consequences of the operation remain to be determined.

The vast majority of children with hypoplastic left heart syndrome—another rare abnormality that consists of a severely underdeveloped left heart, including the ventricle and ascending aorta—die within the first week after birth. Two recent surgical approaches have had encouraging results, however. One is the Norwood procedure, which does not provide a cure but does allow children to grow enough to undergo a Fontan-type operation. The Norwood procedure creates a new outflow for the heart, using the pulmonary artery, and creates a shunt to provide blood flow to the lungs.

The other approach to these babies is heart transplant shortly after birth. Early results are encouraging, but the long-term effects of the necessary drugs (which repress the immune system and fight rejection) on growth and development in children are not well understood. However, with these two approaches, there is some hope for these infants who otherwise cannot survive.

Two other more common congenital problems, easily treated surgically, are patent ductus arteriosis and coarctation of the aorta. A patent ductus arteriosis can be treated with a simple surgical procedure that does not require open-heart surgery. The ductus arteriosis is a blood vessel that connects the aorta and the pulmonary artery while the child is still in the womb. Its purpose is to allow blood to bypass the lungs, because they are not functioning before birth. Blood flows from the right side of the heart to the pulmonary artery directly to the aorta and out to the body. In a few cases, this vessel fails to close normally within a few days after birth and remains open or “patent.” Blood then flows into the lungs and may cause heart failure. The defect usually can be treated.
HEART SURGERY

successfully in the newborn period with medicine or a simple surgical procedure in which the abnormal connection is closed with a small metal clip, applied through an incision in the left side of the chest.

Coarctation of the aorta refers to a congenital narrowing of the aorta after it leaves the heart. This obstructs and reduces blood flow to the rest of the aorta, the arms, legs, kidneys, and other vital organs. This condition also causes an increase in blood pressure near the blockage (in the arms). The constriction is removed by surgical reconstruction of the aorta. Normal circulation is restored. High blood pressure, which may become permanent, is common in patients who have not been treated early enough, but when it is adequately treated in childhood, the hypertension is most often relieved eventually. Some patients may have a “recurrence” of coarctation later in life. This can often be repaired by a balloon dilation procedure done in the cardiac catheterization laboratory. (See Chapter 24.)

Children with coarctation usually have no symptoms early in life. An elevated blood pressure, heart murmur, diminished pulses in the legs, or poor development of the lower extremities may suggest the diagnosis.

SPECIAL CONSIDERATIONS IN PEDIATRIC HEART SURGERY

Like all heart surgery, this area is highly specialized and should be performed only in medical centers by surgeons who devote a large part of their professional careers to the treatment of congenital heart disease. Availability of experts in pediatric cardiology, pediatric anesthesia, and pediatric intensive care is essential to the successful outcome of these operations. Technological advances in all these areas have led to lower mortality and better long-term success rates.