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Drs. Alfred Blalock (left) and Helen Taussig (right).

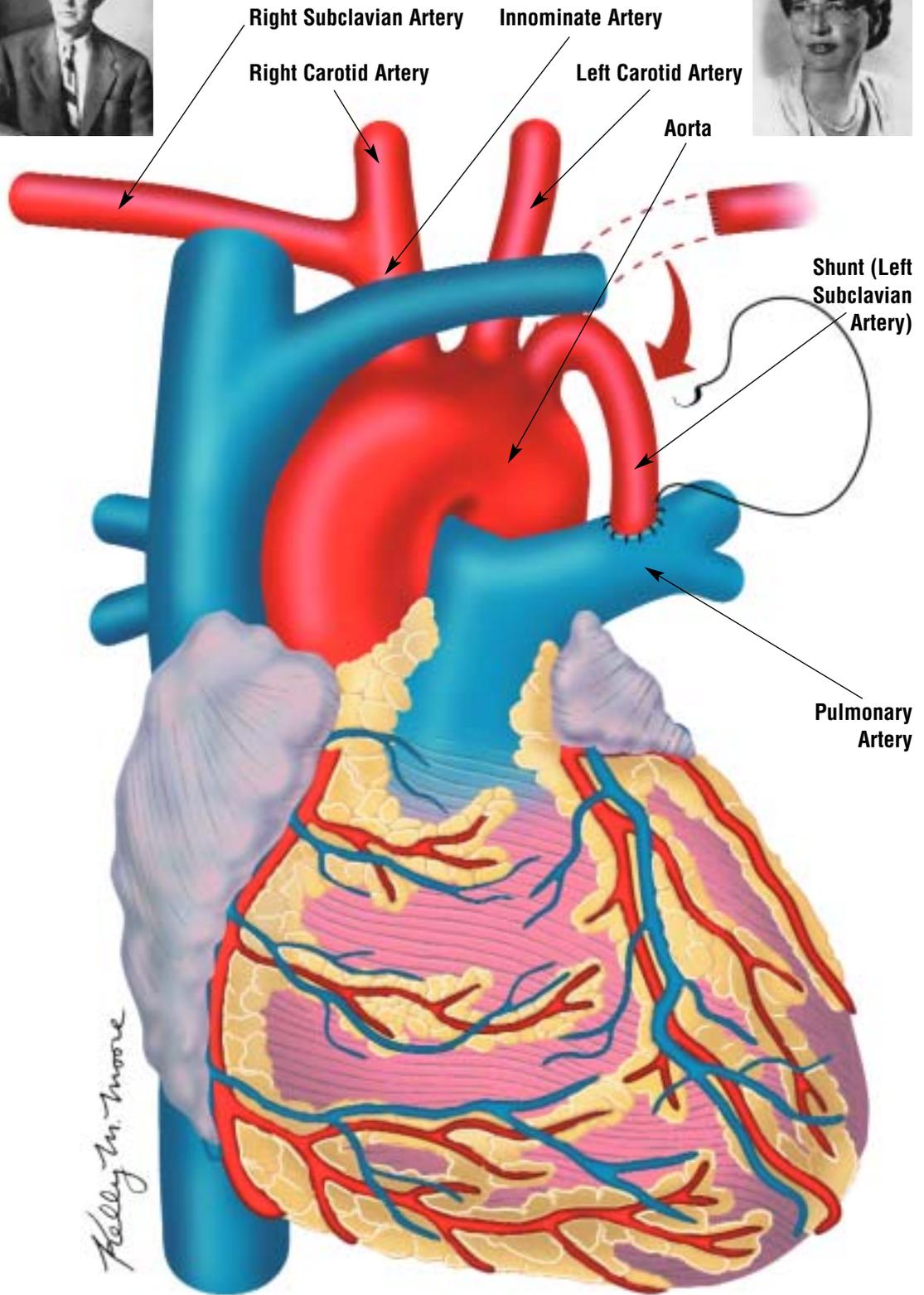


Fig. 7.1: The famous “blue baby operation,” or Blalock-Taussig shunt, was the first surgical procedure developed to treat a congenital heart defect. In this operation, an artery from the arm is connected to the pulmonary artery to help supplement blood flow to the lungs and thus provide more oxygenated blood to the body.

Fig. 7.1

HEART PROBLEMS OF INFANTS AND CHILDREN

 **A** CONGENITAL HEART DEFECT means an abnormality that is present at birth. Congenital heart surgery started before the heart-lung machine was developed as surgeons started to work on abnormalities of the arteries that came out of the heart. Dr. John Streider at the Massachusetts General Hospital in Boston tied off a patent ductus arteriosus (an abnormal pathway between the aorta and the pulmonary artery) in a child on March 6, 1937. Unfortunately, the patient was quite sick at the time and died four days after the surgery. A year and a half later, in the same city, on August 16, 1938, Dr. Robert Gross at the Boston Children's Hospital operated on a girl seven and a half years old who was short of breath because of the same congenital defect. The patient made a successful recovery. Soon surgeons all over the world were performing this operation.

The next major congenital cardiovascular defect to be overcome was called "coarctation of the aorta." This is a defect in which the aorta narrows and, in some cases, becomes totally blocked, resulting in decreased blood flow to the lower half of the body. Historically, most patients with this defect eventually died of complications by the age of twenty years. In 1944, Dr.

Clarence Crafoord in Stockholm, Sweden, successfully removed this narrow area of the aorta in a twelve-year-old boy.

Only a year later, Gross successfully treated a condition known as vascular ring, which occurs when the aorta and its major arterial branches wrap around the esophagus and trachea in an abnormal manner and compress them. Before this landmark surgical procedure, many infants and children with the condition died of suffocation and/or starvation.

During that same period, doctors announced the famous "blue baby operation" at Johns Hopkins University Hospital in Baltimore. This operation treats congenital heart defects in which the unoxygenated blood returning from the body is shunted through a hole in the heart instead of going through the lungs and is pumped out through the aorta, which causes the child to have a bluish color.

The first patient was a fifteen-month-old girl who had suffered her first cyanotic spell (turning blue) at age eight months. Dr. Helen Taussig, a pediatric cardiologist at Johns Hopkins University Hospital, took her as a patient and consulted with other doctors about trying a new operation. Over the next several months, the baby girl refused most of her



Dr. Robert Gross performed the world's first successful surgical closure of a patent ductus arteriosus, an abnormal pathway between the aorta and pulmonary artery in newborns.

Table 7-1: First intracardiac repairs using heart-lung machine or cross circulation

Congenital Heart Defect	Year	Surgeon	Comment
Atrial septal defect	1953	Gibbon	Heart-lung machine (HLM)
Ventricular septal defect	1954	Lillehei	Cross circulation
Complete atrioventricular canal	1954	Lillehei	Cross circulation
Tetralogy of Fallot	1954	Lillehei	Cross circulation
Tetralogy of Fallot	1955	Kirklin	HLM
Total anomalous pulmonary veins	1956	Kirklin	
Congenital aneurysm, sinus of Valsalva	1956	Kirklin	
Congenital aortic stenosis	1956	Kirklin	First directly viewed correction
Aortopulmonary window	1957	Cooley	First closure using HLM
Double outlet right ventricle	1957	Kirklin	Extemporaneously devised correction
Corrected transposition of great arteries	1957	Lillehei	
Transposition of great arteries: atrial switch	1959	Senning	Physiologic total correction
Coronary arterial-venous fistula	1959	Swan	
Ebstein's anomaly	1964	Hardy	Repair of atrialized tricuspid valve
Tetralogy with pulmonary atresia	1966	Ross	Aortic allograft
Truncus arteriosus	1967	McGoon	Aortic allograft
Tricuspid atresia	1968	Fontan	Physiologic correction
Single ventricle	1970	Horiuchi	
Subaortic tunnel stenosis	1975	Konno	
Transposition of great arteries: atrial switch	1975	Jatene	Anatomic correction
Hypoplastic left heart syndrome	1983	Norwood	Two-stage operation
Pediatric heart transplantation	1985	Bailey	

Table 7-1:

Within a few years in the mid- to late 1950s, surgeons corrected thirteen types of congenital heart defects. Congenital heart surgery later evolved into its own subspecialty.

feedings and lost weight. She weighed only 8.3 pounds at the time the operation was performed by Dr. Alfred Blalock at Johns Hopkins University Hospital on November 29, 1944. During the operation, Blalock sewed an artery that normally supplies blood to the arm to the left pulmonary artery so more blood could get to the lungs and be oxygenated (Fig. 7.1). The successful operation required slightly less than an hour and a half. Although this was not a cure for her heart condition, it improved the patient's symptoms and quality of life substantially.

Thus, within a seven year period, three congenital cardiovascular defects — patent ductus arteriosus, coarctation of the aorta, and vascular ring — were all attacked surgically and treated successfully. However, the introduction of the Blalock-Taussig shunt was probably a much more powerful stimulus to the development of open heart surgery because the operation **palliated** a complex intracardiac defect and focused attention on the abnormal physiology of cardiac disease.

Palliative:

A treatment that improves a condition but does not cure it.

The next major step forward in heart surgery needed to wait for the development of the heart-lung machine, which occurred in the middle 1950s. With the advent of techniques to support the circulation and oxygenate the blood, using either the cross circulation technique of Dr. C. Walton Lillehei or the modified Gibbon-IBM heart-lung machine of Dr. John Kirklin, the cardiac teams of the University of Minnesota and the Mayo Clinic led the way and did many of the first intracardiac repairs for a number of commonly occurring congenital heart defects. Palliative operations, however, continued to be used and developed to improve circulatory physiology without directly addressing the anatomic pathology. The palliative operations somewhat improved the patients' conditions but did not cure them. As the safety of the heart-lung machine steadily improved, surgeons addressed more and more complex congenital abnormalities of the heart in younger and younger patients. Some of the milestones in the development of op-

erations to correct congenital defects using cardiopulmonary bypass appear in Table 7-1.

Diagnosing a Congenital Heart Defect

The human heart begins to develop at the end of the first month of fetal life and takes about another eight weeks before it resembles an adult heart. During this period, about eight out of every one thousand newborns develop some form of congenital heart defect ranging from very mild to quite severe. The exact cause of congenital heart defects is unknown, but recent information suggests there may be genetic influences. In some cases, they are associated with other medical conditions, such as the mother contracting German measles (rubella) while pregnant. At this point, most doctors don't think congenital heart defects are hereditary in the strict sense of being passed from parent to offspring, but children of parents who were born with such a defect will be somewhat more likely to have a congenital heart defect.

Some congenital heart defects are diagnosed shortly after birth or even while the baby is in the uterus by using ultrasound or echocardiography. They

may be diagnosed later when the child is of school age, or in rare circumstances, the congenital cardiac defect remains hidden until adulthood. One indicator of some types of congenital heart defect in a newborn is a faint bluish color in the skin. Some children with heart defects may not thrive, and many suffer from congested lungs, which may be related to heart failure. Heart murmurs can also indicate congenital heart defects, although not necessarily.

If a defect in a newborn is suspected, your child's pediatrician will recommend an electrocardiogram and probably an echocardiogram, which do not require any needle sticks. Other tests used to diagnose congenital heart defects include cardiac catheterization and magnetic resonance imaging. After the heart defect is diagnosed and analyzed, your pediatrician and pediatric cardiologist will develop a treatment plan. This may require nothing more than yearly checkups or perhaps medications. Occasionally, a catheter can be used to dilate a heart valve or to insert a plug to close a hole. Heart surgery may be recommended and, in rare cases, heart transplantation is the best option.

Specific Defects

There are many types of congenital heart defects. Of the following eleven congenital heart defects, the first nine are relatively common. The last two are much more rare and included for a sense of perspective on the challenges facing a congenital heart surgeon. I also have purely personal reasons for mentioning them. In the 1970s and early 1980s, I was fortunate enough to work with Dr. C. Everett Koop at the Children's Hospital of Philadelphia. It was my privilege to have been involved with the care and surgery of some of these patients. Koop, later to become surgeon general of the United States, was then chief of pediatric surgery and

Table 7-2: Relative frequency of occurrence of cardiac malformations at birth

Disease	Percentage
Ventricular septal defect	30.5
Atrial septal defect	9.8
Patent ductus arteriosus	9.7
Pulmonary stenosis	6.9
Coarctation of the aorta	6.8
Aortic stenosis	6.1
Tetralogy of Fallot	5.8
Complete transposition of great arteries	4.2
Truncus arteriosus	2.2
Tricuspid atresia	1.3
All others	16.5

Source: *Heart Disease: A Textbook of Cardiovascular Medicine.*

Table 7-2: This table shows the most common congenital heart and major vessel defects. The ventricular septal defect, comprising almost 30 percent of congenital heart defects, is by far the most common.

had cared for patients with both types of these very rare and difficult defects.

Ventricular Septal Defect

In this most common congenital defect there is a hole in the septum that separates the right and left ventricles (Fig. 7.2). As a result, blood is short-circuited back into the lungs, putting a burden on both heart and lungs. About 30 percent to 50 percent of these holes, especially the smaller ones, close over time. Patients with large- or moderate-size defects that do not close spontaneously, however, eventually need an operation to close them. Larger defects may have to be closed in the first year of life because they can cause shortness of breath and other symptoms of heart failure. If the defect is not closed, the patient can also develop pulmonary vascular disease, which damages blood vessels in the lungs and can eventually be fatal.

Ventricular septal defects vary in size and location, so naturally some of them are more easily closed surgically than others. While in surgery, the patient's heart and lung function are provided by a heart-lung machine, and the actual hole is closed

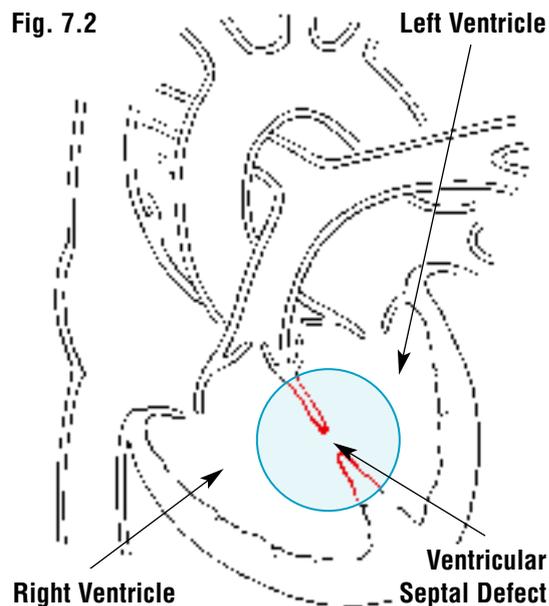


Fig. 7.2

Ductus Arteriosus:
A tube connecting the pulmonary artery to the aorta. After birth, when the lungs begin to function, this tube normally closes.

Fig. 7.2: Ventricular Septal Defect:
In this defect there is a hole in the wall of muscle, or septum, that separates the left and right ventricles. It is usually corrected with a patch.

with a patch. The chances of surviving the surgery in childhood and subsequently living a normal life are superb.

Patent Ductus Arteriosus

While the fetal heart is developing, a tube develops between the aorta and the pulmonary artery. This tube, called the **ductus arteriosus**, is responsible for bypassing the lungs, moving blood from the pulmonary artery to the aorta. Because the fetus receives oxygenated blood from its mother through the placenta, it has no need for functioning lungs. After the child is born, however, the lungs begin to function, and the ductus arteriosus is no longer needed. It normally closes from a couple of hours to a couple of days after birth. However, if it remains open, or patent, it is considered a congenital defect and usually needs treatment (Fig. 7.3).

In some cases, the patent ductus arteriosus is so large that enough blood is shunted back from the aorta into the low-pressure pulmonary artery to actually flood the lungs. Heart failure can develop because the heart is working so hard to pump blood, and much of it is just being short-circuited back to the lungs. In other cases, infections develop in the tube, or over time high blood pressure in the pulmonary arteries can result in what's called pulmonary vascular disease. This disease damages the blood vessels in the lungs where resistance to blood flow increases and, at some point, the blood flow can actually reverse. If this happens, the right ventricle, which should be pumping un-oxygenated blood into the lungs, is actually pumping un-oxygenated blood through the ductus directly into the aorta. This condition causes blueness, or cyanosis, and is very serious. Fortunately, the open ductus is usually diagnosed well before this condition develops, and the defect can be corrected.

There are three ways to close a patent ductus. In newborn babies, especially

SIR BARRATT-BOYES: OPENING HEART SURGERY “DOWN UNDER”

WHILE TRAINING TO BECOME a heart surgeon, native New Zealander Sir Brian Barratt-Boyes had the opportunity to spend two years working at the Mayo Clinic.

“I was assigned to various surgeons,” he said. “Dr. John Kirklin was one. He had recently begun there, but he was making a name for himself, and in 1955, began doing open heart surgery with the Gibbon-IBM heart-lung machine. It was a staggering responsibility.

“We went through a very rapid learning curve,” Barratt-Boyes said. “Some of it was pretty traumatic, but the results were exceptional. I used to be sent as a spy to Minneapolis to see what Dr. C. Walton Lillehei was doing. I reported what was happening so I saw the cross circulation operations as well, which were very fascinating.”

In 1956, he returned to his native New Zealand and entered a kind of creative vacuum. New Zealand was isolated from the medical community. Later, in fact, Barratt-Boyes would be credited with founding modern open heart surgery in the Pacific Rim area.

“If you are working independently and follow your own star and your own ideas, you can sometimes come up with something that wouldn’t be

the case if you worked in a group, where everybody has their own ideas,” Barratt-Boyes said.

It was there that Barratt-Boyes made two contributions to cardiac surgery. In 1962, only a month after Dr. Donald Ross in England, Barratt-

Boyes performed the world’s second successful aortic valve **homograft** implantation. “It worked extremely well, even in the first patient, who was a young girl of about fifteen with **endocarditis**,” Barratt-Boyes said. “She had a successful operation and recovered from her endocarditis. She’s now had several children and works outside the home. She is a remarkable person.”



Brian B. Barratt-Boyes

After this, Barratt-Boyes, working with surgeons from Japan, helped introduce and popularize open heart surgery in infants by using hypothermia, or lowering the body temperature to protect the brain.

His results with infants, often desperately ill and with complex forms of congenital heart disease, set new world standards. This method was an important stepping stone, and even today most centers use variations of hypothermia techniques in some infants and adult patients.

Homograft:

A donor graft, or piece of tissue, taken from a donor and placed into a recipient of the same species.

Endocarditis:

An infection involving the heart, caused by bacteria or virus.

Fig. 7.3: Patent Ductus Arteriosus:

An abnormality in which a tube connects the pulmonary artery to the aorta, mixing unoxygenated and oxygenated blood. This tube is open during fetal development when the lungs are not needed but is supposed to close after birth.

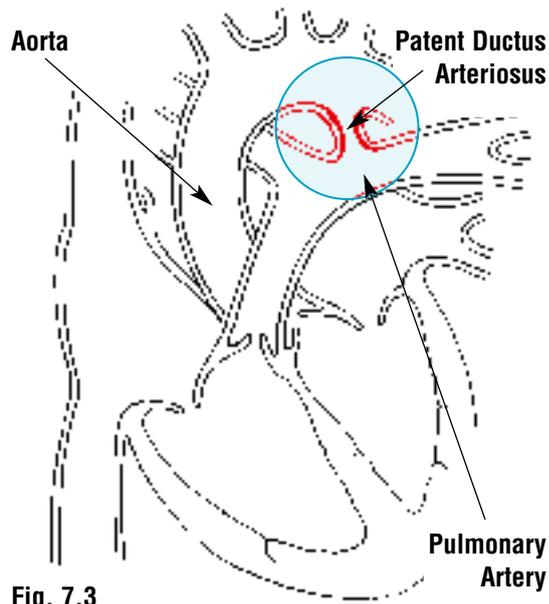


Fig. 7.3

premature newborns, it can frequently be closed by giving a medicine called indomethacin, which causes the ductus to constrict and close. This treatment does not always work, however.

The conventional treatment is surgical closure. This is done by opening the chest on the left side and dividing the ductus and oversewing its ends, or closing it with a tie or a metal clip.

There are also catheters that can be threaded through blood vessels to deliver devices that actually plug the ductus. This avoids a surgical incision in the chest. This procedure has advantages and disadvantages that should be discussed with the pediatrician and the pediatric cardiologist. Trial tests of these devices are being evaluated by the U.S. Food and Drug Administration (FDA).

With all methods, the chances of surviving the closure procedure are better than 99 percent, and in most cases the patient is cured.

Atrial Septal Defect

Fig. 7.4: Atrial Septal Defect:

An abnormal opening in the wall of muscle, or septum, that divides the two filling chambers, or atria, of the heart.

An atrial septal defect (ASD) is a hole in the common wall separating the two atria (Fig. 7.4). There are different types of atrial septal defects. In most cases, they are

well tolerated by children and may not be diagnosed until the child is older — and sometimes not even until adulthood.

Once diagnosed, however, most of these should be closed. Depending on the size, the hole can be sewn up. Larger holes may require a patch of pericardial tissue or an artificial material such as Dacron.

If the hole is not repaired, heart failure can develop because much of the blood pumped by the heart is being short-circuited through the lungs instead of being pumped out to the body, meaning the heart has to work harder to pump more blood. Occasionally, pulmonary vascular disease may develop, or blood clots dislodged from veins in the legs may travel through the ASD and lodge in the brain, causing a stroke. In most cases, the risk of the surgery to repair the defect in children is low, and the survival rate is greater than 99 percent. Devices to close ASDs with a catheter are under development and are being tested at some centers.

There is another, more complex atrial septal defect that may occur with a ventricular septal defect called atrioventricular canal defect. The risks associated with the surgical repair of this defect

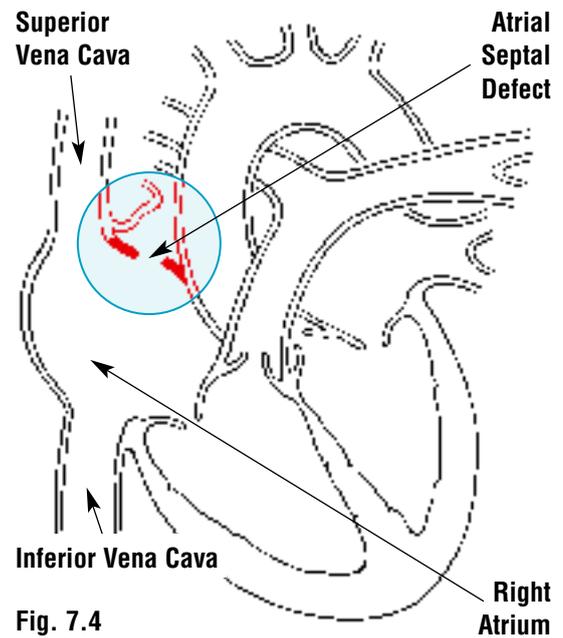


Fig. 7.4

are somewhat higher. Children with Down's syndrome have a higher chance of having atrioventricular canal defect.

Any surgical repair of atrial septal defect requires a heart-lung machine, and afterwards most patients can look forward to a normal life expectancy.

Coarctation of the Aorta

In coarctation of the aorta, there is an abnormal narrowing of a short segment of the aorta, usually less than an inch long (Fig. 7.5). The aorta can be narrowed up to 90 percent in this area. Over time, it can become totally occluded. If the aorta is narrowed, blood to the lower body bypasses the narrowing by using collaterals, or tiny channels. In a healthy person, these collaterals are barely functioning, but in patients with coarctation they can become very large.

This defect can be diagnosed at birth or shortly afterwards. Typical signs include high blood pressure in the arms and abnormally low blood pressure in the legs, and strong pulses in the arms and minimal or absent pulses in the legs. Some infants may develop severe heart failure, and an emergency operation may be required. In less severe cases, coarctation of the aorta is not diagnosed until a child is older and sometimes not until he or she is a teenager or an adult.

The life span of people with coarctation of the aorta can be severely shortened if they do not have surgical correction. This is partially because of the high blood pressure in the upper body, which can result in strokes or heart failure. Also, infection is prone to occur at the point of the coarctation, or the aorta can rupture near the coarctation.

In surgery, doctors can remove the narrowed area and suture the normal ends of the aorta back together. Or they can widen the narrowed area with a patch, or replace the narrowed area with a tube made of Dacron. This is a curative

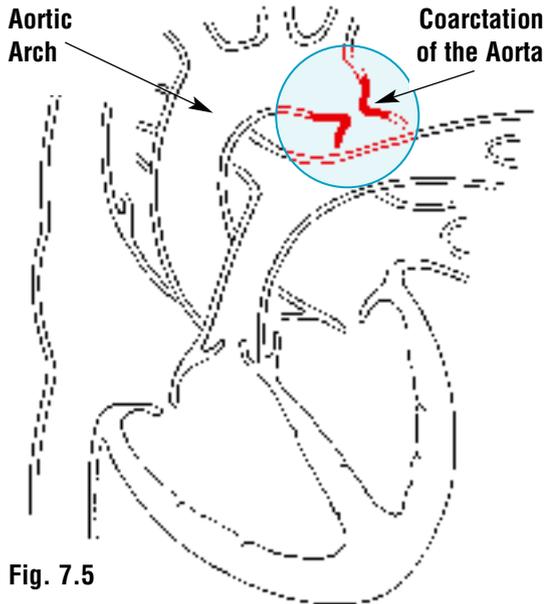


Fig. 7.5

operation. The chance of surviving repair of isolated coarctation of the aorta is greater than 99 percent. The arteries that supply blood to the spinal cord sometimes originate from the aorta in the area of the coarctation. Because of this, about one in two hundred patients undergoing surgical repair develops some degree of paralysis of the lower half of the body. Occasionally the defect can recur and has to be reoperated on. If it does recur, the narrowed segment can sometimes be dilated with a balloon catheter.

Transposition of the Great Arteries

In transposition of the great arteries, the two main arteries coming out of the heart, the aorta and the pulmonary artery, are switched (Fig. 7.6). As a result, when the unoxygenated blood returns from the veins into the heart, it is pumped directly back into the aorta, making the infant very cyanotic. The oxygenated blood that returns from the lungs is pumped back into the lungs through the pulmonary artery, which branches off the left ventricle. When children have any congenital heart defect in which the child is cyanotic or the

Fig. 7.5: **Coarctation of the Aorta:**
An abnormal narrowing of the aorta after it leaves the heart.

skin is a bluish color, they are sometimes referred to as “blue babies.” In some cases of transposition of the great arteries, there also may be other heart defects, such as ventricular septal defect.

Infants suffering from transposition are usually quite cyanotic, and if it is not corrected, they will probably not survive their first year of life. Before the advent of the heart-lung machine, there were some palliative operations developed, but these procedures were not cures. In fact, even today many infants with transposition undergo a procedure to make or enlarge a hole in the atrial septum shortly after birth. This procedure is done in the cardiac catheterization laboratory with a special catheter threaded up through a blood vessel in the groin. The hole in the atrial septum allows mixing of blood and temporarily improves the infant’s condition until a surgical repair can be made.

Currently there are a number of ways this defect can be repaired surgically. One technique involves switching the pulmonary artery back to the right ventricle and the aorta back to the left ventricle. This procedure is technically challenging because the coronary arteries that come

off the aorta are quite small, particularly in infants. These tiny coronary arteries have to be moved and reconnected to the aorta in its new location.

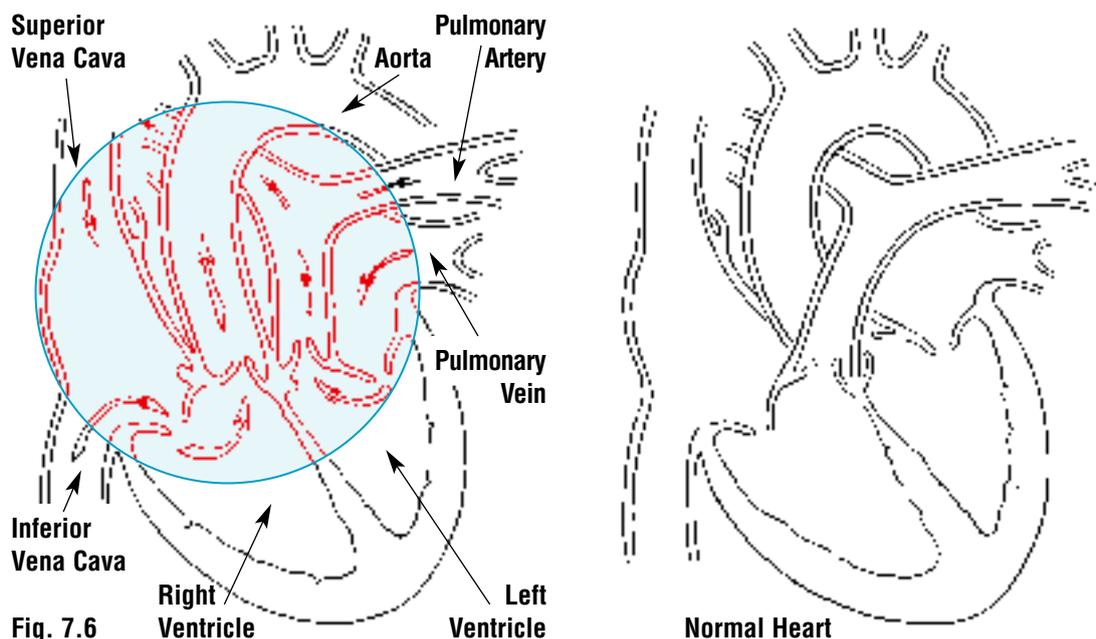
With current surgical techniques, the chances of surviving the surgical procedure, depending on the complexity of the transposition malformation, are between 90 and 95 percent. The long-term survival is good for most patients.

Tetralogy of Fallot

This complicated condition is actually four different congenital defects occurring simultaneously in the same heart. Tetralogy, in fact, means “set of four,” and Fallot was a French physician who was one of the first to describe the condition.

In this defect (Fig. 7.7), 1) there is a ventricular septal defect; 2) blood flow from the right ventricle to the pulmonary arteries is obstructed (the obstruction can be at the pulmonary valve or in the right ventricular outflow tract leading to the valve and/or in the pulmonary arteries themselves); 3) the aortic valve overrides the ventricular septal defect; and 4) the right ventricle is abnormally thickened.

Fig. 7.6: Transposition of the Great Arteries: When the two main arteries arising from the heart, the pulmonary artery and the aorta, are switched, causing unoxygenated blood to be pumped back out through the aorta into the circulation. The normal heart at far right is shown here for comparison.



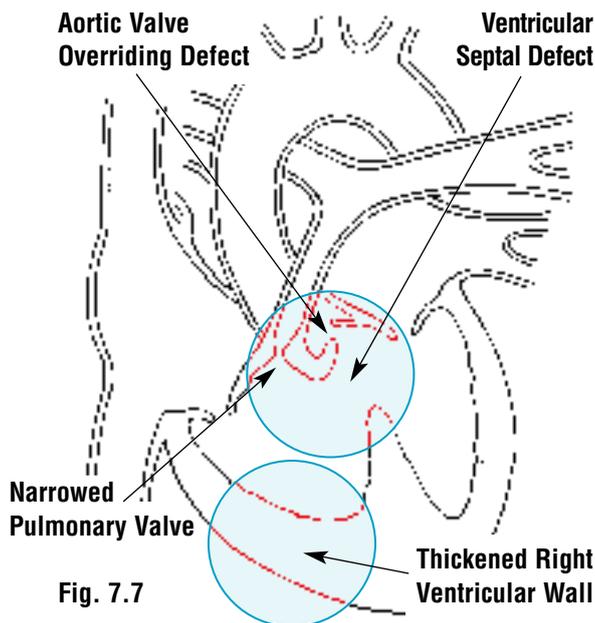


Fig. 7.7

Because of the location of the ventricular septal defect in relation to the aortic valve and the obstruction of blood flow through the pulmonary arteries to the lungs, unoxygenated blood returning from the body is shunted from the right ventricle to the left ventricle. It mixes with the oxygenated blood returning from the lungs, which results in blueness or cyanosis. In some forms of tetralogy of Fallot, the defects are more severe, particularly in terms of the amount of unoxygenated blood flowing from the right ventricle across to the left ventricle. The more severe the obstruction, the more cyanotic the patient. In some cases, newborns will suffer from severe cyanosis and may have to undergo urgent surgery.

In most cases, doctors recommend that the defect be corrected sooner rather than later, usually within the first six months of life. The surgical repair includes closing the ventricular septal defect and relieving the obstruction of blood flow to the pulmonary arteries. In some special circumstances, however, palliative shunting procedures are recommended before a complete repair is made.

Because of the cyanosis associated with this defect, patients used to be called

“blue babies,” and the early surgical shunts, like the Blalock-Taussig shunt, that were performed were known as “blue baby operations.” These operations would alleviate a good deal of the cyanosis and restore the children to a more normal color.

The chance of surviving a primary surgical repair is greater than 90 percent. Long-term survival in most of the patients whose defect is repaired is good.

If the defect is not repaired, serious complications can develop, including progressive cyanosis, strokes and infections of the brain, pulmonary hemorrhage, and severe hypoxic spells related to a lack of oxygen. Without surgical intervention, most patients with tetralogy of Fallot will not survive until their twentieth birthday.

Pulmonary Valve Stenosis

Pulmonary valve stenosis is a narrowing of the heart valve located between the right ventricle and the pulmonary artery (Fig. 7.8). When the valve is very narrow, the patient may have substantial symptoms while still an infant, including a bluish tinge to the skin. This defect can be life threatening. In older children, symp-

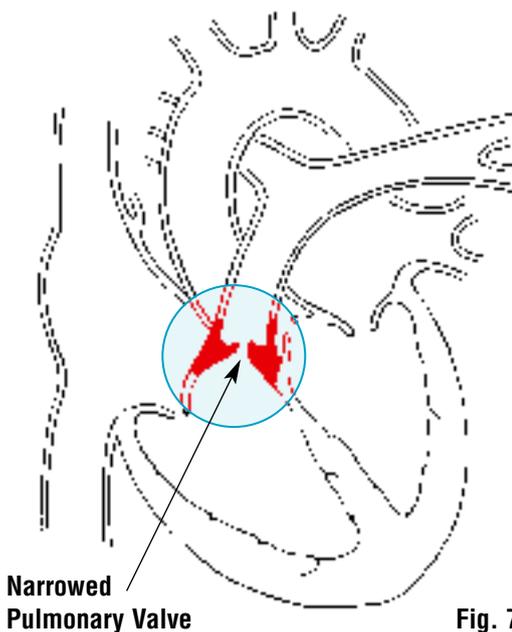


Fig. 7.8

Fig. 7.7:

Tetralogy of Fallot:

A set of four individual defects, including: 1) a ventricular septal defect; 2) an obstruction of blood flow from the right ventricle to the pulmonary arteries; 3) overriding of the aortic valve above the ventricular septal defect; and 4) an abnormally thickened right ventricle.

Fig. 7.8: Pulmonary Valve Stenosis:

An abnormally narrowed pulmonary valve, which is located between the right ventricle and the pulmonary artery.

Heart Murmur:

A noise produced from blood flowing through the heart or other blood vessels or through the lungs.

toms include fatigue or a reduced ability to exercise. The child may stand out because he or she cannot keep up with the other children physically.

The diagnosis is often suspected after hearing a **heart murmur** and obtaining an electrocardiogram. An echocardiogram will likely identify the defect and allow a definitive diagnosis to be made. For simple pulmonary valve stenosis, a balloon catheter is used to dilate the valve, usually with good results. Sometimes surgery using the heart-lung machine is necessary. In some cases, there is also muscular obstruction within the right ventricle, and this tissue needs to be removed. The results, after balloon dilatation or surgery, are usually excellent. The chance of surviving these procedures is greater than 99 percent. The long-term results are usually excellent.

Congenital Aortic Stenosis

In this defect's simplest form, the aortic valve is abnormally narrowed (Fig. 7.9). Other variations of this defect include narrowing of the aorta immediately above the heart valve or a membrane obstructing blood flow below the aortic valve. In some

cases, an abnormality of the muscle immediately below the heart valve causes obstruction. Children with severe forms of aortic stenosis are likely to have symptoms of heart failure and shortness of breath. This defect may be suspected because of a heart murmur or because of an abnormal ECG. Confirmation of the diagnosis would be made with an echocardiogram. In some instances a cardiac catheterization may be necessary to make the diagnosis.

With the most severe forms, an infant may require emergency heart surgery to open the valve. Some pediatric heart centers use a balloon catheter to open the narrowed heart valve, thus postponing valve surgery until the child is older.

The decision to recommend heart valve surgery depends on how serious the obstruction is. This defect may be so mild that surgical intervention is unnecessary. If surgery is necessary in straightforward and uncomplicated forms of the disease, the chance of surviving the procedure is greater than 98 percent. The chance of surviving the surgery is somewhat lower in more complex forms of the disease, and surgery is higher risk in critically ill **neonates**. With a successful procedure, long-term results are often very good, although in many instances, a second heart valve operation may be needed later.

Neonate:

A newborn, or a child within the first several weeks after birth.

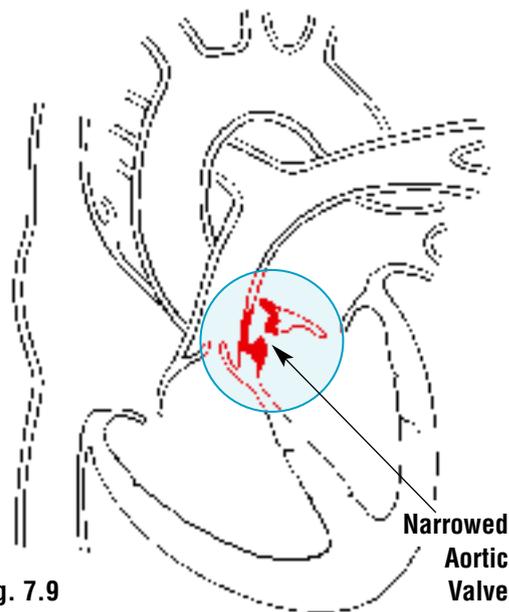


Fig. 7.9

Fig. 7.9: Congenital Aortic Stenosis:

This defect is usually an abnormal narrowing of the aortic valve, which is located between the aorta and the left ventricle. There are, however, several different forms of this defect.

Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome (HLHS) is one of the most severe and life-threatening malformations of the human heart. "Hypoplastic" means "underdeveloped," left heart refers to the structures that make up the left side of the heart, and syndrome means a group of things that appear together.

The job of the left heart is to receive oxygenated blood from the lungs and distribute it to the body. The left heart includes the left atrium (left filling chamber), the left ventricle (main pumping

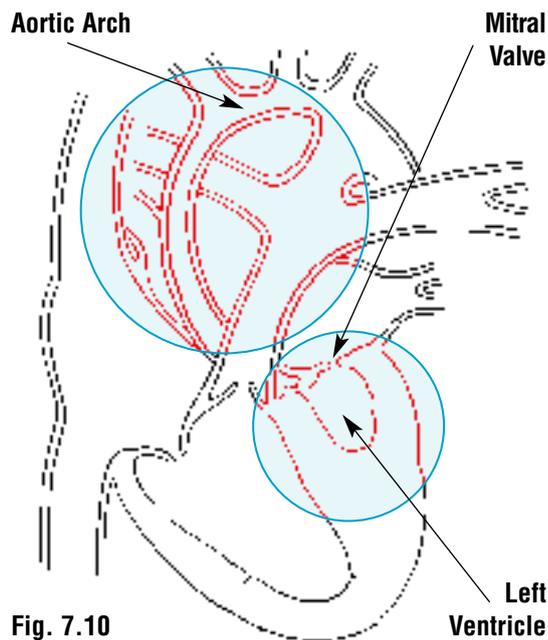


Fig. 7.10

chamber), the mitral valve (valve between the left atrium and the left ventricle), the aortic valve (valve between the left ventricle and the aorta), and the aorta. HLHS consists of a wide spectrum of malformations in which one or more of these structures are critically small (Fig. 7.10). Despite the variation that can exist in cases of HLHS, the net result is the same: The left side of the heart cannot do its job properly.

Unfortunately, this is not a rare condition. Data from the New England Regional Infant Cardiac Program found HLHS to be present at a rate of 0.163 per one thousand live births.

Without surgery, 99 percent of patients with HLHS will die shortly after birth. In 1983, Dr. William Norwood at Boston Children's Hospital reported that he had successfully operated on an infant in two separate stages about a year and a half apart. That two-stage operation has greatly improved the chance of survival for infants born with HLHS. Today, Norwood's two-stage repair has evolved into a three-stage repair performed over the first few years of the patient's life.

The goal of these three operations is to bypass the small left side of the heart by making the right ventricle the heart's main pumping chamber. Since the right ventricle will, therefore, no longer be available to perform its usual job of pumping blood to the lungs, the vessels that normally carry the unoxygenated blood (superior and inferior vena cavae) are connected directly to the lung arteries. The flow of unoxygenated blood to the lungs occurs passively without the benefit of an intervening pumping chamber. This type of passive pulmonary blood flow in a heart with only one good pumping chamber is called a Fontan operation — another variation is the Glenn operation. They are applicable not only in HLHS but also in other types of abnormal hearts with a single pumping chamber.

The advantage of the three-stage Norwood procedure is that these operations can almost always be performed; the disadvantage is that it requires three operations and still results in a heart with only one pumping chamber. Furthermore, the long-term results of using the right ventricle (instead of the left ventricle) to pump oxygenated blood to the body are not known.

Dr. Leonard Bailey at Loma Linda University has pioneered the use of newborn heart transplantation for the treatment of HLHS. If a donor heart is available, cardiac transplantation requires only one operation and produces a structurally normal heart with two pumping chambers. Unfortunately, about 25 percent of the newborns who would need a cardiac transplantation die of complications while waiting for a donor heart. After cardiac transplantation, the patient must take antirejection medication for the rest of his or her life. The side effects of these medicines can become serious over time, and rejection of the transplanted heart can occur. Incidentally, Bailey created quite an uproar in the news media in 1984 when a human heart donor was not

Fig. 7.10: Hypoplastic Left Heart Syndrome: This literally means underdeveloped left heart. Some or all of the structures on the left side of the heart, including the heart's main pumping chamber, are undersized. It is a very serious congenital defect.

available and he transplanted the heart of a baboon into a desperately ill twelve-day-old premature girl known as "Baby Fay," who was suffering from HLHS.

At this point it is not clear which surgical treatment is better for hypoplastic left heart syndrome. Both, however, are an improvement on previously available treatments. HLHS used to be 100 percent fatal during the first year of life. Although pediatric heart surgeons tend to hold strong opinions, neither the Norwood procedure nor cardiac transplantation is a perfect treatment option for HLHS, and neither approach is clearly superior. In the best centers, roughly 60 percent of patients who undergo either surgery are alive ten years later.

Ectopia Cordis

This is a rare congenital heart defect. As of 1989, only 219 cases had ever been reported in the medical literature.

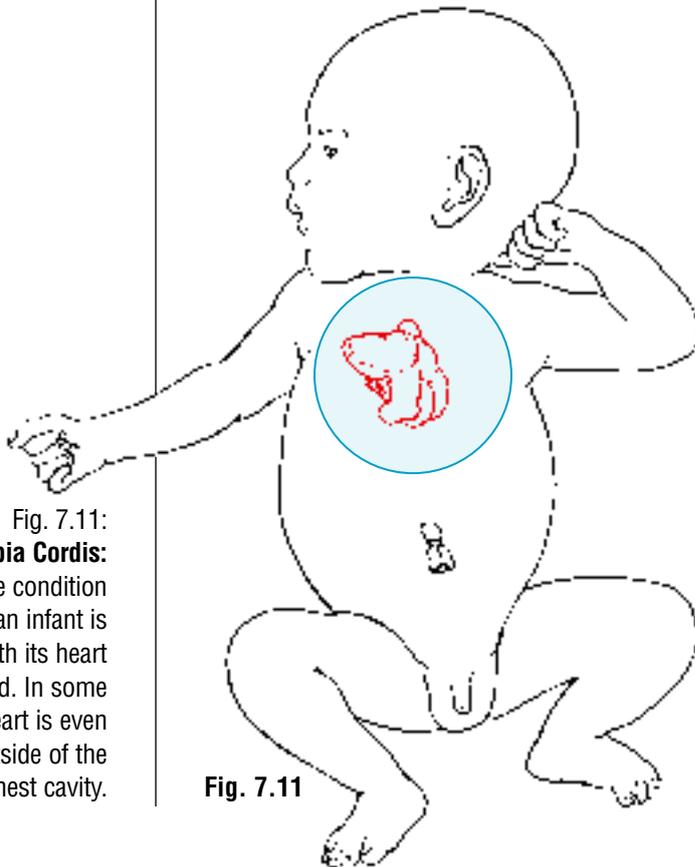


Fig. 7.11:
Ectopia Cordis:
A rare condition in which an infant is born with its heart mislocated. In some cases, the heart is even located outside of the chest cavity.

Fig. 7.11

Ectopia means displacement, and cordis means heart.

There are four types of this defect: the heart may be located in the neck, either partially or completely in the abdomen, or, almost unbelievably, outside of the body on the chest (Fig. 7.11). Many infants suffering from this defect have other abnormalities inside the heart as well.

When the heart is outside of the body, it is recommended that the infant undergo immediate surgery to put the heart back into the chest. The first successful surgery of this type was performed by Dr. Narish Saxena at the Children's Hospital of Philadelphia in 1975. This case, in which I was involved, was that of a child who had to undergo multiple innovative surgeries and practically lived the first few years of his life in the hospital.

Thoracopagus Twins

Identical twins occur at an incidence of four per one thousand births. Conjoined twins are identical twins who are joined to each other in some place on their bodies. This form of twin is much rarer and has an incidence of one per fifty thousand to one hundred thousand births. In 1811, in the country of Siam (now Thailand), a Chinese mother gave birth to identical twins joined at the hip. The boys were named Eng and Chang, and they lived unseparated until their death at age sixty-three years. They became famous as a result of being promoted as "freaks" by P.T. Barnum, who called them the "Siamese Twins," and the term has been used since then for all twins who are born attached to each other.

Most Siamese twins are born dead, but there are about four hundred sets known to have lived, ranging in life span from just a few hours to the sixty-three years of Eng and Chang. They can be joined in various places on the body, including the lower back, the abdomen, the hip, the leg, and even the head. About 40

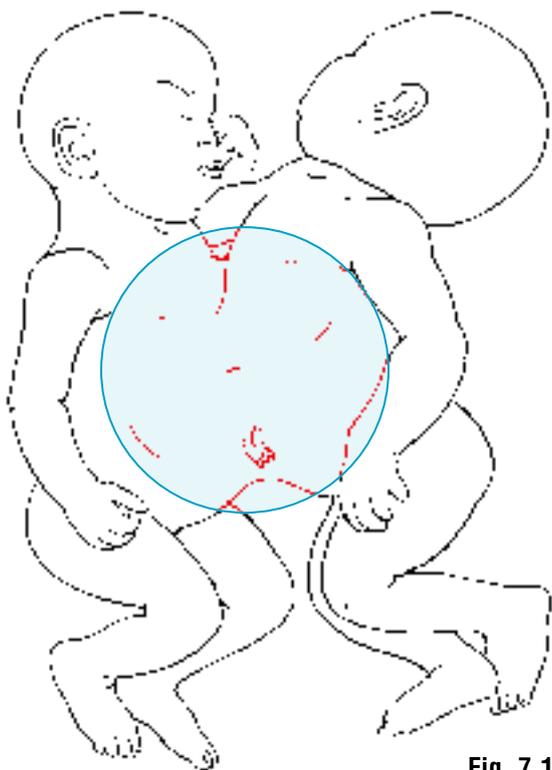


Fig. 7.12

percent of the surviving twins are joined at the chest, face to face, looking each other in the eye (Fig. 7.12). They are called “tho-

racopagus twins.” (Thoraco refers to the chest; pagus is a Greek word meaning fixed). Thoracopagus twins may also be joined at the abdomen down to the pubis. They may share a common liver, intestines or even a heart.

For pediatric heart and thoracic surgeons, the division of thoracopagus children ranges from extremely challenging to outright impossible with today’s knowledge. My experience in one case with Koop, the chief pediatric surgeon, and Dr. L. Henry Edmunds, Jr., the chief heart surgeon at the University of Pennsylvania, involved a set of Siamese twins with only one heart between them. Before the surgery could be done, the children had to be taken to the operating room and the procedure mapped out ahead of time, position by position, turning them in different ways for each incision. The surgery was considered a success even though we were able to save only one child, but generally success in this very complicated field has been limited. Twin separation remains a challenge for the pediatric surgical community.

Fig. 7.12:
Conjoined Twins:
 Commonly referred to as “Siamese twins,” these are a set of twins who are joined at some place on their bodies, including at the chest.