

Therapeutic Cardiac Catheterizations for Children with Congenital Heart Disease

Introduction

A therapeutic cardiac catheterization is a procedure performed to treat your child's heart defect. A doctor will use special techniques and a thin plastic spaghetti-like tube or catheter that goes to the heart from blood vessels in the legs or the neck. These techniques allow the repair to be done without surgically opening the chest and heart. The types of defects that can be repaired include closing a hole in the wall that separates the heart's right and left sides, widening a narrowed vessel or stiff valve, and closing abnormal blood vessels using a variety of devices.

There are different ways in which your child will be kept comfortable during his or her cardiac catheterization. These depend on

- your child's age,
- diagnosis,
- type of intervention to be performed, and
- a variety of other factors,

Your doctor will decide whether your child will receive sedation only or general anesthesia. Before entering the cardiac catheterization laboratory (cath lab), a medication may be given to help your child relax and fall asleep (figure 1).

If sedation is chosen, your child will also have an IV started. This allows medications to be given to keep your child asleep and pain-free during the procedure. Your child will keep breathing on his own, and no breathing tube will be needed. Sometimes a topical numbing cream may be applied to both an IV site and to the groin area where the catheters will be placed.

If your doctor chooses general anesthesia as best for your child, you will most likely meet with an anesthesiologist prior to the catheterization. Either an inhaled gas via a mask or an intravenous medication may be used for general anesthesia. After your child is asleep, a breathing tube is inserted into the airway to make breathing easier. Additional medications and fluids may be given throughout the procedure to keep your child comfortable. When the procedure is over, the breathing tube will be removed when your child is breathing on his own.

Catheterization is a sterile procedure performed using small catheters placed into blood vessels, so the risk of infection is minimal. Antibiotics aren't usually needed before or after the procedure. Removing the catheter, however, may cause blood to ooze into the skin. This can discolor the skin, like a black eye, but it doesn't usually cause problems. No treatment is required.

The catheter doesn't hurt the heart and it isn't painful once inside the heart, but its movement can cause abnormal heart rhythms. Your doctor can usually treat these rhythms by removing the catheter or using medications. Rarely, if the catheter touches the heart's electrical system it can interfere with the spread of electricity. This is known as heart block. Although this is usually temporary, placing a special catheter connected to an electrical battery (pacemaker) may be required until the heart's electrical system corrects itself.

The Cardiac Catheterization Procedure

After your child is asleep, the part of the body where the catheters will be inserted is prepared (or "prepped") by washing the skin with an antiseptic solution to ensure that it's clean. Your child is positioned for safety and comfort. Additional sedation may be given as needed. Sterile drapes will cover your child, the table and some of the equipment used during the procedure.

During the therapeutic cardiac catheterization, specialized tubes called sheaths are placed into the blood vessels in the groin or neck area. This allows the doctor to get catheters through these vessels and into the heart. In the newborn period, the "belly-button" vessels (the umbilical vessels) may be used for access to the heart. A local anesthetic is generally used before placing catheters and sheaths in the body.

The movement of these catheters as they pass from one heart chamber to the next is carefully monitored with X-ray images on a TV screen in the cath lab. Through the catheters, pressures are measured and samples of blood are drawn



Figure 1: Example of a cardiac catheterization laboratory. Large monitors are used to see the heart and the catheters magnified under X-ray.

from various sites within the heart. This gives the doctor valuable information about your child's heart function and the blood flow through the heart and lungs. Contrast (X-ray dye) is injected through the catheters to get a clear movie picture of the internal heart structures to identify where the defect is and how severe it is. This information helps your doctor determine the best way to treat the defect.

In the following pages, various types of therapeutic cardiac catheterizations are covered. Your doctor will discuss the risks and benefits of the procedure.

When the procedure is complete, all catheters and sheaths are removed. This is similar to removing an IV. For several minutes pressure is put on the spot where the catheters were inserted. To prevent bleeding, a pressure dressing may be applied.

Your child is then transferred to a stretcher and taken to the recovery area.

Therapeutic Cardiac Catheterization

In the last two decades, tremendous medical advances have been made. These allow congenital heart defects to be treated in the cath lab without surgery.

Valvuloplasty or Balloon Valvotomy

This procedure is done to open a narrowed heart valve. Any of the heart's four valves can be narrowed. However, this procedure is most often used to open the valves connecting the heart to the lungs (pulmonary valve) or to the body (aortic valve). These narrowings occur because the valve leaflets don't open up completely. This makes it harder for the heart to pump blood to the lungs or to the body (figure 2a). The narrower the valve, the more pressure it takes to pump the blood through it. It's like asking someone to breathe through a straw. The narrower the straw, the harder it is to breathe. To open the narrowed valve, a special catheter with a balloon attached to its end is used.

This catheter is advanced from a blood vessel in the groin or neck through the narrowed valve. Then the balloon is inflated to widen the opening.

A picture of the valve is first taken and the size of the valve is measured carefully to select the correct-size balloon. If the balloon is too small, the opening may still not be big enough. If the balloon is too large, the valve may be damaged or the vessel may be torn. The balloon is inflated for only a few seconds, then it is deflated and removed (figure 2b).

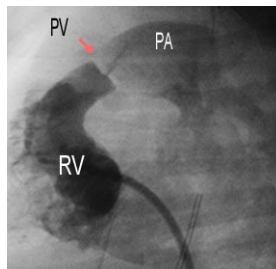


Figure 2a: Picture in the right ventricle (RV) looking from the side shows severe obstruction to flow through the pulmonary valve (PV), with only a pin-hole size opening into the pulmonary artery (PA).

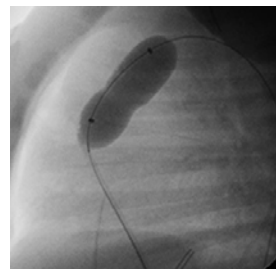


Figure 2b: A balloon is positioned through the valve and inflated to widen the opening.

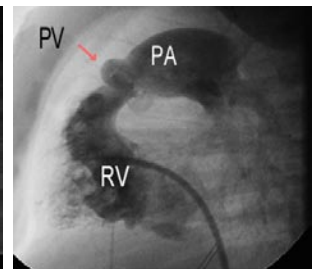


Figure 2c: The balloon is removed and a picture in the right ventricle (RV) afterwards shows a wide opening at the pulmonary valve (PV).

Pressures in the heart and pictures of the valve are repeated to assess the results (figure 2c).

Angioplasty

This procedure widens a narrowed blood vessel. These narrowings are often associated with various congenital heart defects and can occur naturally or after surgery. Similar to a narrowed valve, a narrowed vessel restricts blood flow and causes the heart to work harder. Blood vessels that can be narrowed include

- the branch pulmonary arteries (vessels that send blood from the heart to the lungs),
- the aorta (major vessel that sends blood from the heart to the body),
- systemic veins (vessels that bring blood from the body back to the heart) and
- pulmonary veins (vessels that bring blood from the lungs back to the heart) (figure 3a).

The procedure is similar to a valvuloplasty in that a balloon catheter is placed inside the narrowed blood vessel and expanded to stretch open the vessel (figure 3b).

Stent Implantation

Sometimes, simply widening a narrowed blood vessel with a balloon isn't effective. The narrowing in the vessel may be too long or it might stretch out with the balloon but shrink again once the balloon is removed.

In this situation, a stent is used to provide structural support within the narrowed vessel to keep it wide open. Stents are metal mesh tubes. They're designed to stretch open inside a narrowed blood vessel and hold the vessel wall open (figure 4).

There are many types of stents, but the most common ones used in children are "balloon-expandable" stents. These are mounted onto a balloon and positioned at the site of narrowing through a long sheath. Then the balloon is inflated to expand the stent against the narrowed vessel wall (figures 5a, 5b, 5c, 6a, 6b). The stent is opened to the appropriate size depending on the patient's size. Then balloon is deflated and removed while the stent stays in the vessel to keep it from renarrowing. Your doctor will carefully measure the narrowed vessel to select the most appropriate-size stent and balloon.

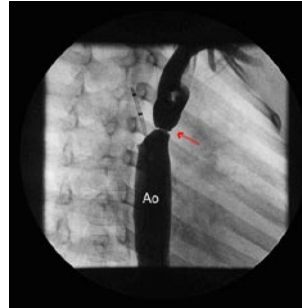


Figure 3a: Picture in the aorta (Ao) shows tight narrowing (red arrow). This condition is called coarctation of the aorta.

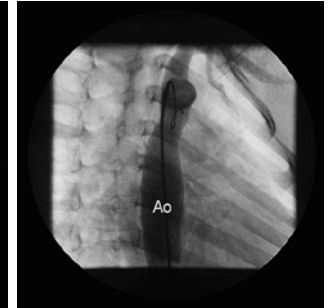


Figure 3b: Picture in the aorta (Ao) shows marked improvement after balloon inflation.



Figure 4: Example of a stent.

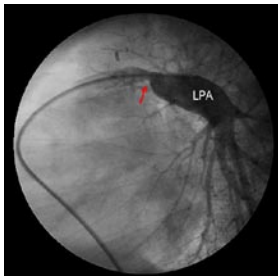


Figure 5a: A picture in the left pulmonary artery (LPA) shows a narrowing (arrow).

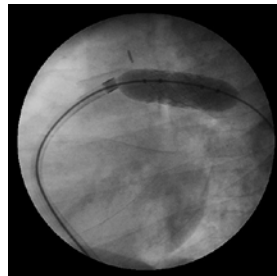


Figure 5b: A balloon is inflated to expand the stent across the narrowing.

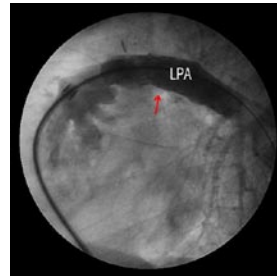


Figure 5c: The balloon is removed and a picture after placing the stent (arrow) shows that the narrowing in the left pulmonary artery (LPA) has resolved.



Figure 6a: A picture in the aorta (AO) shows a tight narrowing (arrow) called coarctation of the aorta.



Figure 6b: After stent placement the narrowing is resolved (arrow).

After stent implantation, your child will usually be placed on a blood-thinning medicine, such as aspirin. This helps prevent clots from forming while the blood vessel wall heals over the stent during the next several months. As your child grows bigger, he or she can be brought back to the cath lab and the stents can be further widened to accommodate growth to adult size. Once implanted, stents can't be removed except by surgery.

Implanted stents don't activate metal detectors. Magnets and microwaves don't affect the stent or the surrounding heart tissue and blood vessels.

Balloon and Blade Septostomy

In some special circumstances, it's necessary to create a larger hole between the walls of the heart's upper chambers (the right and left atrium). Special balloons and blade catheters are used to create these openings to increase blood flow between the heart's upper chambers. This procedure can be performed in the cath lab or by the bedside in the intensive care unit under ultrasound guidance.

Valve Perforation

Some patients are born with a completely blocked pulmonary valve. This is called pulmonary atresia. When this occurs and blockage is due to a thin membrane of the valve, the blocked valve can be opened in the cardiac cath lab using radiofrequency perforation. This technique uses a special catheter that can generate heat to create a small opening in the blocked valve. Then the catheter can be placed across the valve and the opening enlarged using the same technique as described in the Valvuloplasty section. When the blockage is due to a thickened abnormal valve, it may not be feasible to make an opening and surgery might be needed.

Occlusion Procedures

These procedures are used to plug up (or close) an unwanted opening or connection in the heart or in blood vessels. These heart defects can be closed using a variety of devices:

1. Secundum Atrial Septal Defect (ASD)

This is a defect in the wall separating the heart's upper chambers. If this opening exists, blood flows abnormally from the left side to the right side, causing the right heart to work harder and less efficiently. Over many years, the pressure in the right heart increases, the heart enlarges and abnormal rhythms can occur. The pressures in the lungs can increase too, and eventually right heart failure can develop.

A catheter device can be used to close this defect (figure 7). The first step in closing an ASD is to measure the size of the defect with pictures taken inside the heart and with ultrasound images taken with a probe placed either down the throat (transesophageal echocardiogram in figure 8) or within the heart (intracardiac echocardiogram). A special sizing balloon is also used to measure the "stretched diameter" of the defect so that the most appropriate size device is selected to provide the best fit.

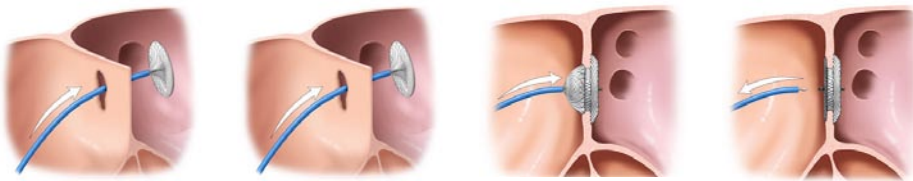


Figure 7: (Courtesy of AGA Medical Corporation) The 4 drawings illustrate closure of an atrial septal defect using the Amplatzer Septal Occluder. First, the left atrial disc of the device is opened within the left atrium. Then, the device is brought back towards the septum so that the disc will lay against the septum. The right atrial disc of the device is then formed, so that the septum sits between both discs, with the device now closing the hole. Once the device is confirmed to be in good position, it is released.

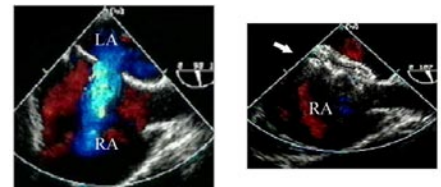


Figure 8: Example of an atrial septal defect as seen by transesophageal echocardiogram prior to device closure (left hand side), and following device closure (right hand side). In the first image the hole is shown between the left atrium (LA) and right atrium (RA), with flow (blue) from LA to RA across the defect. In the second image, a device is seen in good position, closing the hole effectively, as no flow can now be seen across the defect.

The device is delivered through a long sheath placed from a blood vessel in the groin. It's positioned using X-ray and ultrasound images. Once it's properly positioned, it's released. Pictures are repeated to check for complete sealing.

Occasionally, a small leak across the defect is noted right after implant. This usually seals off over the next few weeks to months. This procedure is commonly performed on an outpatient basis or with an overnight hospital stay.

Patients are usually kept on a mild blood thinner such as aspirin for about six months while the body heals over the device.

A listing of current FDA-approved devices is included in the addendum.

2. Patent Foramen Ovale (PFO)

This is a small, naturally occurring opening in the wall separating the heart's upper chambers. This hole is present in all newborns; by adulthood it should have sealed shut. In 10–25 percent of people, the opening remains unsealed and blood can flow from the heart's right to left side under special circumstances.

There are no symptoms associated with a PFO. It's not a heart defect, and doesn't increase the heart's workload. However, in rare circumstances it can be a source of a stroke. If a blood clot forms in the veins, the clot can travel to the right side of the heart and cross this hole to the left side, then go into the brain to cause a stroke. Currently there are no medical indications to close a PFO just because it exists. But if a PFO is found after a stroke, it may be recommended to have the opening closed. The benefits and risks of closing a PFO in stroke patients is being studied.

3. Fenestrated Fontan

This is a unique circumstance reserved for patients with complex congenital heart disease involving a single ventricle. The Fontan baffle or conduit is surgically placed to direct blood flow from the body to the lungs. The fenestration is a small hole in the Fontan baffle that the surgeon strategically creates to allow a small amount of blood to flow from the Fontan baffle directly to the left heart without going through the lungs (figure 9a, 9b). This is generally thought to benefit the patient, especially early after surgery.

Once the patient has recovered from the operation, your cardiologist may feel the fenestration is no longer needed and should be closed.

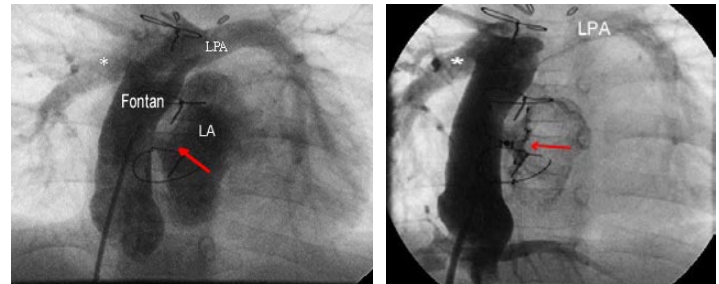


Figure 9a: Picture in the Fontan baffle (Fontan) shows a fenestration (arrow), through which low oxygen blood flows to the left atrium (LA), lowering the oxygen levels in the body.

Figure 9b: Immediately after closure with a device (arrow) there is less flow through the opening. It is expected that healing will take place and close the hole completely. *Right Pulmonary Artery; LPA: Left Pulmonary Artery; LA: Left Atrium.

4. Ventricular Septal Defects (VSD)

This is a defect in the wall separating the heart's two lower chambers (ventricles). Blood flows abnormally from the (higher pressure) left side to the (lower pressure) right side through the defect. This causes the entire heart to work harder and less efficiently. In general, compared to an ASD of the same size, significantly more blood flow crosses a VSD. This is because the pressure difference between the two ventricles is much more than the difference between the two atria.

Since the right heart sends the extra blood flow to the lungs, these get congested due to the extra volume of blood going through them. The amount of blood flow also depends on the size of the defect.

Many defects will get smaller on their own or even close in the first two years of life and may never need treatment. However, the larger defects typically need to be closed because if they aren't, they cause heart failure. If large defects are left untreated over many years, the pressures in the right heart and in the lungs will increase, the heart will become enlarged and thickened, and a severe condition called pulmonary hypertension may occur.

Surgery is still the most common way to close these defects. Depending on the shape and location of the VSD, few can actually be closed without surgery using a device. In particular, some types of muscular VSDs can be closed in the cardiac catheterization laboratory with a procedure similar to that of closing an ASD (figures 10a, 10b).

X-ray images and a transesophageal or intracardiac echocardiogram are used to guide the delivery of the device. A special sizing balloon can be used to measure the "stretched diameter" of the defect so the most appropriate size device is selected for the best fit. The device is delivered through a long sheath placed from a blood vessel either in the neck or groin, depending on the exact location of the VSD. Once the device is in proper position, it's released. Just as in ASD closures, patients are kept on mild blood thinner such as aspirin for about six months while the body heals over the device.

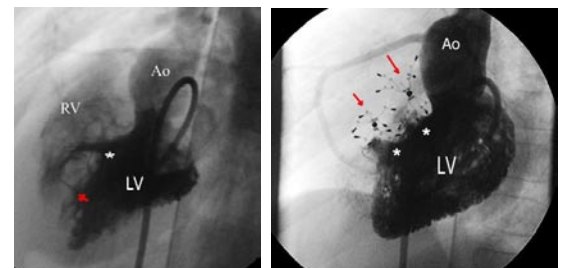


Figure 10a: Picture in the left ventricle (LV, red dotted area) shows multiple holes in the septum or ventricular septal defect (arrow and *). These holes allow for blood to go from the left ventricle to the right ventricle (RV, blue dotted area), instead of going to the aorta (Ao).

Figure 10b: Two devices were implanted (arrows). The picture in the left ventricle (LV) a year later shows no significant flow left through the holes (*). The right ventricle no longer fills from the left ventricle.

5. Patent Ductus Arteriosus (PDA)

This is a naturally occurring blood vessel that connects the major blood vessel leaving the heart and going to the lungs (the pulmonary artery) and major blood vessel leaving the heart and going to the body (the aorta). This vessel is present in all newborn infants; it should close within a few days after birth. However, in some patients, this vessel remains open and shunts blood away from the aorta to the pulmonary artery. This can lead to heart failure and/or risk a heart infection during a lifetime. X-ray pictures of the PDA are taken and its dimension is measured carefully to select the appropriate-size device to implant (figures 11a, 11b, 11c).

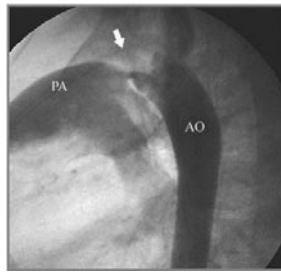


Figure 11a: Picture in aorta (Ao) as seen from the side shows an open PDA (arrow), with blood flowing from the aorta to the pulmonary artery (PA).

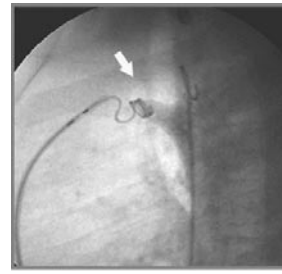


Figure 11b: A coil is implanted in the PDA (arrow).

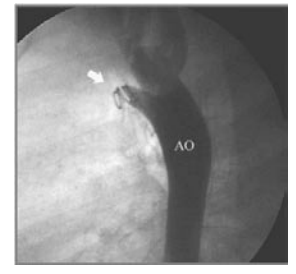


Figure 11c: After delivery of the coil, a picture in the aorta shows that the coil closed the PDA completely, and there is therefore no further blood flow from the aorta to the pulmonary artery.

This device is delivered through a long sheath from a blood vessel in the groin. Once the device is in a stable position within the PDA, it's released (figure 12a, 12b). Your child won't need to take additional medications such as aspirin after this procedure.

People with these implanted devices can still use microwave ovens or magnets. The devices aren't affected by metal detectors or MRI scans.

Other Closure Procedures

Other abnormal blood vessels occasionally need to be plugged. They include the vessels listed below. For the most part, coils are used.

1. Aorto-Pulmonary Collaterals

These are abnormal blood vessels that go from the aorta (the large artery that supplies oxygenated blood to the body) to the pulmonary arteries (which are blood vessels that take used blood from the heart back to the lungs for oxygen). These collateral vessels cause the heart to work harder that it should. In certain situations they need to be eliminated.

2. Venous Collaterals

These abnormal vessels form off veins. (Veins are blood vessels that return used blood or oxygen poor blood from the body back to the heart.) and go to the left heart or into blood vessels that bring oxygen-rich blood back to the left heart. As a result, some oxygen-poor blood gets into the body and mixes with the oxygen-rich blood. This gives a patient's skin a bluish appearance. These collaterals are mostly found in patients after surgery for a single ventricle.

3. Arterio-Venous Malformations

Arteries are vessels that bring oxygenated blood to the body. Veins are vessels that return oxygen-depleted blood back to the heart. Arterio-venous malformations are generally a network of abnormal blood vessels that communicate between arteries and veins. These abnormal vessels cause the heart to work harder than it needs to. They should be eliminated when possible.

After Catheterization

When your child arrives in the recovery area, he or she will be monitored closely. If your child was put to sleep using general anesthesia, the breathing tube will be removed as soon as your child is breathing on his own. Your child will probably be sleepy for the next few hours. It's important to let your child lie quietly to prevent bleeding at the catheterization site. A pressure bandage may be applied to the site.

After your child wakes up, small amounts of clear liquids are offered at first. This will grade into a regular diet as tolerated. If nausea or vomiting occurs, the oral fluids may be continued for a period of time before your child tries to eat again. Occasionally, a medicine to prevent further nausea and vomiting may be given.

The IV will be removed before discharge.

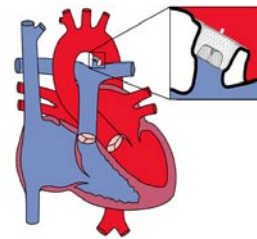


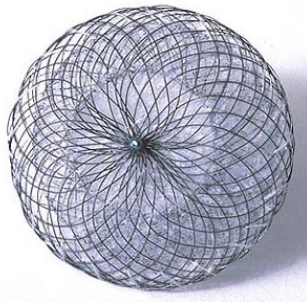
Figure 12a: (Courtesy of AGA Medical Corporation) This graph illustrates a PDA being closed by a device occluder.



Figure 12b: This picture in aorta (Ao) shows a PDA closed with a device occluder (arrow). There is no residual flow across the PDA.

Addendum

FDA approved devices:

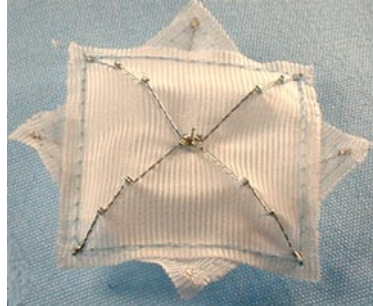


Courtesy of AGA Medical Corporation

Figure 13: Photograph of an Amplatzer Septal Occluder as seen from the front (A) or from the side while pulling from each end (B).



Courtesy of AGA Medical Corporation



Courtesy of NMT Medical Corporation

Figure 14: Photograph of the CardioSEAL Septal Occluder, as seen from the front.



Courtesy of AGA Medical Corporation

Figure 15: Picture illustrates an Amplatzer Duct Occluder, attached to the delivery catheter.

Discharge

After many therapeutic cardiac catheterizations, patients can be discharged later that same day. Sometimes an overnight hospital stay is required. Your doctor will decide when your child can go home.

Before your child leaves to go home, your doctor or nurse will give you instructions on how to take care of the dressing and the insertion site. Generally, this site should be kept clean and dry and watched for signs of infection such as redness, swelling, tenderness and drainage. There may be bruising and mild discomfort where the catheter was inserted, but this usually goes away in the next few days. It's not uncommon to have a low-grade fever for the first 24 hours after a catheterization. You'll be given instructions about how to take care of the fever or discomfort, when to bathe or swim, and what kinds of activities to avoid. For infants, the diaper should be kept dry for the first few days after the procedure to avoid contamination at the catheter insertion site.

If you have any questions or concerns about your child's health, feel free to contact your cardiologist.

ASD

The Amplatzer Septal Occluder is an FDA-approved device for ASD occlusion (figure 13; also refer to figures 7 and 8).

This device looks like two disks woven together at its center. It's made out of Nitinol, a nickel-titanium alloy. This metal can be bent out of shape (such as by being squeezed into a tiny catheter) and then, when it's released, it will spring back to its original shape. Inside the two disks are patches of Dacron sewn on for enhanced occlusion. The Gore Helex Septal Occluder is another device recently approved for ASD closure. The device is made of a patch material supported by a flexible nitinol wire with circular shape. Another device for ASD occlusion undergoing investigational trials is the STARFlex Occluder.

PFO

Devices available for this procedure, which have been approved by the FDA under Humanitarian Device Exemption regulations, include: The Amplatzer PFO Occluder and the CardioSEAL Septal Occluder (figure 14).

The Amplatzer PFO Occluder is similar to the other Amplatzer devices. The difference is the shape. The center is thinner and the right disk is slightly larger than the left one.

The CardioSEAL is also a double disk made of a Dacron patch sewn onto a stainless steel frame. It looks like two umbrellas facing each other and attached at the center. The device can be folded down to fit into a long thin tube for delivery. The technique of delivery is similar to that of ASD closures.

Fenestrated Fontan

Devices available for closing the fenestration include the CardioSEAL (figure 14; also refer to figures 9a and 9b) and the Amplatzer Septal occluders. Again, the techniques are similar to those used to close a PFO or ASD.

VSD

The CardioSEAL Septal Occluder (figure 14; also refer to figures 10a and 10b) is an FDA-approved device for closing muscular VSDs.

Other devices under investigation include the Amplatzer VSD Occluder and the STARFlex device.

PDA

A variety of metallic coils are used to plug a PDA. The most commonly used one is the Gianturco coil. It's made of flexible loops of stainless steel and has a mesh of Dacron fibers attached to it to enhance occlusion. Various techniques have been developed to ensure the coils are safely placed. Your cardiologist will discuss these with you.

Sometimes, the PDA can be quite large and tube-like. These can be plugged with a "sack of coils" known as the Gianturco-Grifka sack. The Amplatzer Duct Occluder (figure 15) is a device approved by the FDA. It's been found to be very effective in closing larger PDAs. This is manufactured similarly to the Amplatzer ASD Occluder but it's more plug-shaped to provide a better fit for the larger cone or tubular-shaped PDA (see figures 12a and 12b).