Adult Congenital Heart Association

Q&A

Transposition of the Great Arteries after Arterial Switch or Rastelli Procedure

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What is D-Transposition of the Great Arteries (d-TGA)?

Transposition of the great arteries (TGA or d-TGA) is a complex congenital heart defect. The two main arteries, the pulmonary artery and aorta, are reversed. It is more common than the other type of TGA – CCTGA (also known as l-TGA).

Normally, oxygen-poor blood from the body is pumped to the lungs by the pulmonary artery and oxygen-rich blood is pumped to the body by the aorta. In babies born with d-TGA, the opposite happens. Unoxygenated blood coming into the right side of the heart from the body is pumped back to the body through the aorta. Similarly, oxygenated blood entering the left side of the heart from the lungs is pumped back to the lungs through the pulmonary artery. If oxygenated blood can't cross over to the other side of the heart and mix, the organs will not receive enough oxygen to properly function.

How often does it occur and what problems does this cause after birth?

TGA occurs in between 1 in 3,500 and 5,000 births. It is more common in males than females.

Babies born with d-TGA show symptoms at birth or soon afterwards. Whether the baby has a way to mix their blood or not will determine how severe their symptoms are. Some babies are born with connections or holes in the septum (atrial septal defects [ASD], ventricular septal defects [VSD] or patent ductus arteriosus [PDA]). These may provide enough mixing of oxygen-rich and oxygen-poor blood to deliver enough oxygen to the body for the short term. If there is no connection or hole, babies will have severe symptoms, as they have no means to get oxygen-rich blood to their body. At birth, babies with d-TGA will appear cyanotic or blue in color. They might have trouble breathing or feeding.

The diagnosis is commonly confirmed with an echocardiogram (ultrasound of the heart). Babies born with a PDA may be given medications through the IV (prostaglandins). These will help to help keep the ductus arteriosus open prior to surgery. Those born without a septal defect might have to have a hole created in the septum before surgery. This can be done in the catheterization laboratory.

Babies born with d-TGA will need surgery shortly after birth to re-route oxygen-rich blood to the body.

What is an arterial switch? How did it become the repair of choice?

In this operation, the two main arteries are detached. The pulmonary artery is then connected to the right ventricle and the aorta is connected to the left ventricle. This allows oxygen-poor blood from the body to be pumped to the lungs and oxygen-rich blood from the lungs to be pumped to the body.

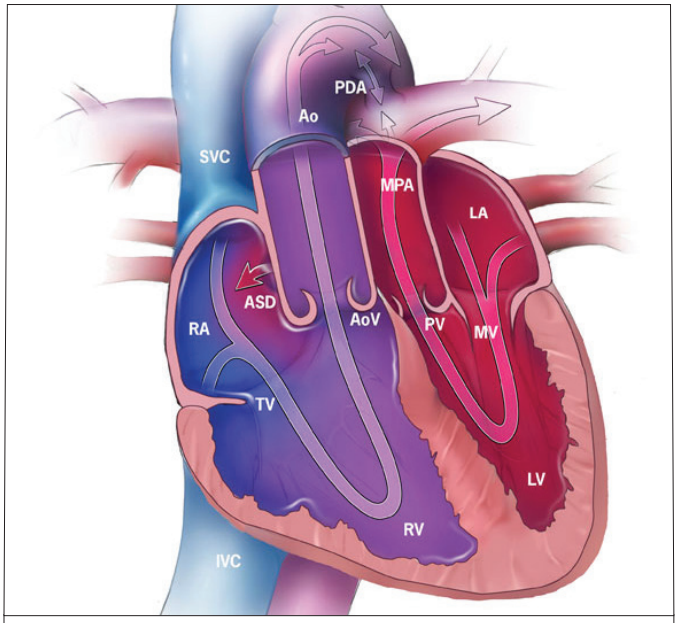
The coronary arteries that supply blood to the heart muscle are removed from the main arteries. They are then reattached to their appropriate positions. Early attempts at arterial switch were unsuccessful. Atrial switches (Mustard repair and Senning repair) were performed instead. Atrial switches eventually resulted in heart failure, and the arterial switch was reintroduced years later.

The arterial switch is now the treatment of choice for simple d-TGA. It preserves the left ventricle as the main pumping chamber for the body. This results in improved long-term outcomes in d-TGA.

What is a Rastelli procedure?

The Rastelli procedure is performed on d-TGA patients with more complex anatomy. It can be used when the main pulmonary artery is too small (pulmonary stenosis) to provide blood flow to the lungs. Because of this, the large arteries cannot be switched.

In this procedure, oxygen-rich blood in the left side of the heart is redirected through a hole between the bottom two chambers of the heart (VSD) to the aorta. This is done by placing a synthetic patch (baffle). The baffle directs blood from the left ventricle to the aorta. To get oxygen-poor blood in the right side of the heart to the lungs, the surgeon also



RA: Right Atrium	SVC: Superior Vena Cava	TV: Tricuspid Valve
RV: Right Ventricle	IVC: Inferior Vena Cava	MV: Mitral Valve
LA: Left Atrium	MPA: Main Pulmonary Artery	AoV: Aortic Valve
LV: Left Ventricle	PDA: Patent Ductus Arteriosus	ASD: Atrial Septal Defect
	Ao: Aorta	

Image of d-TGA courtesy of the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities.

connects a conduit from the right ventricle to pulmonary artery. This is a tube with a valve made of synthetic material, human or animal tissue. This helps bypass the narrowed pulmonary artery. This surgery also allows oxygen-rich blood to travel to the body. Like the arterial switch, it also restores the left ventricle as the systemic ventricle. The Rastelli procedure also has the benefit of not needing to detach and re-attach the coronary arteries.

What problems can occur long-term after these types of repairs?

Both types of repairs have potential problems that require lifelong congenital cardiology follow up.

In the arterial switch patient:

- The coronary arteries must be closely monitored for obstruction. This is more common in patients who have other coronary anomalies.
- Stenosis, or blood vessel narrowing, can also occur in either the pulmonary artery or aorta.
- Aortic dilation has been reported. Blood pressure modification may be helpful in preventing this.
- Aortic regurgitation can also occur.
- Neurocognitive problems are common.
- ADHD, depression and anxiety are often seen.

In a Rastelli patient:

- Repeated conduit dysfunction may occur. This is due to narrowing (stenosis) or leaking (regurgitation) of the conduit. The conduit will eventually need to be replaced. This can sometimes be averted by stenting and/or transcatheter pulmonary valve replacement. Conduits and valves may need to be replaced sooner if they become damaged or infected. Infection can be prevented by taking antibiotics before dental procedures.
- If not followed closely, leaky or narrowed valves and conduits can lead to weakening of the heart muscle and heart failure.
- Additionally, heart rhythm problems can result from the heart surgery and scarring of the heart muscle. Eventually, a pacemaker or defibrillator might be needed.

Is pregnancy safe following arterial switch or Rastelli?

Most women with d-TGA treated with arterial switch or Rastelli tolerate pregnancy and delivery well. However, you should talk to your adult congenital heart disease (ACHD) team before getting pregnant. Your medications may need to be changed. A careful evaluation may also help identify any heart problems that need to be addressed before pregnancy.

You should also talk to an obstetrician in a high-risk pregnancy group. It is important that you keep your appointments with your ACHD doctor and your high-risk pregnancy team during your entire pregnancy. They will keep an eye on your heart function and the health of your baby.

What about exercise? Is it safe for me?

Exercise is important in patients with d-TGA. Regular exercise will keep your heart as strong as possible. While it is generally safe and beneficial, it is important to ask your ACHD doctor before taking on any new physical activity. He/she will tell you which types of activities you can safely do.

How can arterial switch and Rastelli patients improve their long-term outcomes?

Patients with d-TGA should eat heart healthy diets, stay active with regular exercise as permitted, avoid smoking and maintain a healthy weight. Aggressive cardiovascular risk factor modification is essential to prevent the development of coronary artery disease (CAD).

Normally, oxygen-poor blood from the body is pumped to the lungs by the pulmonary artery and oxygen-rich blood is pumped to the body by the aorta. In babies born with d-TGA, the opposite happens.

It is important to regularly follow-up with your ACHD doctor. Catching long-term problems before they cause symptoms is key to preventing permanent heart damage. Patients with an arterial switch or Rastelli may benefit from a daily baby aspirin. This may help decrease your risk for coronary disease and decrease the infection risk in pulmonary artery conduits. Ask your ACHD doctor if this is right for you.

All patients with CHD should take good care of their skin and teeth as these are potential routes for bacteria to get to the heart. This includes seeing your dentist every six months and for some patients, taking antibiotics prior to these visits as instructed by your cardiologist.

What kind of cardiology care is recommended for adults with d-TGA following arterial switch or Rastelli?

The American College of Cardiology and the American Heart Association classify d-TGA as a complex heart defect. This means that experts recommend that you get care for your heart at a special ACHD center. You can find a listing of ACHD clinics at www.achaheart.org/directory.

If you have had an arterial switch or Rastelli, you should see your cardiologist at least every 6-12 months, even if you feel fine. This way you can catch any problems before they start, since new heart problems can develop without symptoms. Many treatments help strengthen and protect your heart if started early. Your doctor will tell you how often to come back. With proper treatment, most babies born with d-TGA grow up to lead healthy, productive lives. It is also important for you to follow up with your primary care provider (PCP) for routine blood pressure, cholesterol and cancer screening. This will increase the chances of living a long and full life.

The good news is that most people with d-TGA and an arterial switch or Rastelli are continuing to do well. By taking care of your heart and getting the recommended care, you can help make sure that you and your heart continue to thrive.

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