

ACHA Q and A: Transposition of the Great Arteries after Mustard/Senning Repair

What is Transposition of the Great Arteries?

In transposition of the great arteries (TGA), the heart's two major arteries are reversed. The blue blood entering the heart gets pumped directly out to the body without going through the lungs. Blood coming from the lungs gets sent back to the lungs rather than out to the body. Babies born with TGA will die unless there is a hole between the two sides of the heart to allow some red blood to get out to the body. Many people with TGA are born with a ventricular septal defect (VSD) that allows this to happen. In others, doctors make a hole to keep them alive. Babies born with TGA almost always need surgery to help the heart work more normally. Two of these surgeries are the Mustard and the Senning repair (also called Mustard or Senning procedures). The main other surgeries are the arterial switch and Rastelli procedure.

What is a Mustard or Senning repair?

The Mustard and Senning repair are two similar operations for TGA. Each is named for the surgeon who first performed the operation. In both, the surgeon creates a two-way baffle in the top part of the heart. This baffle serves as a bridge between the two sides of the heart. In a Senning procedure, the surgeon uses the patient's own tissue to create the baffle. In the Mustard procedure, a synthetic material is used. Both are called "atrial switch procedures" because there is a baffle through the heart's top part, or atria, which allows the blood to reach the ventricles.

When and where were Mustard and Senning repairs first done?

Dr. Ake Senning of Sweden performed the first Senning procedure in 1957. Dr. William Mustard, from Canada, performed the first Mustard procedure in 1963. In general, the Senning was more common in Europe and the Mustard was more common in the United States and Canada.

Are the Mustard and Senning repairs still used?

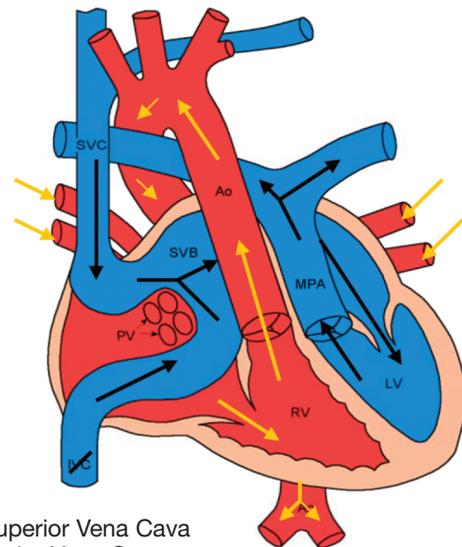
Today's children born with TGA usually have an operation called an arterial switch. This operation was first developed in the late 1980s. In the arterial switch, the surgeon disconnects and reattaches the heart's arteries to create more normal blood flow. This means that Mustard and Senning patients are usually between 22 and 50 years old.

What kind of long-term problems do people with a Mustard or Senning repair experience?

There are three kinds of problems that people with a Mustard or Senning repair face: rhythm problems, baffle problems, and pump problems:

- **Rhythm problems:** Many TGA patients have inborn problems with the heart's electrical system. The scarring from previous surgery can also cause electrical problems. Sometimes the heart's "relay box," called the sinus node, is damaged. This can cause sick sinus syndrome, which makes the heart beat too slowly. Pacemakers are used to treat a too-slow heart. Up to 25% of Mustard/Senning patients have a pacemaker by adulthood. Other TGA patients experience heart rates that are too fast (tachycardia). This happens when a section of the heart's electrical system starts firing too quickly. Heart rhythm doctors now often use ablation to treat too-fast heart rates. The doctor first identifies the area of the heart that is misfiring. Then, this area is scarred to stop the fast heart rate. If the ablation is

In this illustration of a heart with TGA that has had a Mustard/Senning repair, the blue blood flows through the two-way Systemic Venous Baffle (SVB) to the left ventricle. The blue blood is pumped to the lungs to get oxygen. The red oxygenated blood is directed through the Pulmonary Venous (PV) baffle to the right atrium and gets pumped by the right ventricle to the rest of the body.



KEY:

SVC – Superior Vena Cava
 IVC – Inferior Vena Cava
 SVB – Systemic Venous Baffle
 PV – Pulmonary Venous Baffle
 LV – Left Ventricle
 RV – Right Ventricle
 MPA – Main Pulmonary Artery
 Ao – Aorta

Image courtesy of the Columbus Ohio Adult Congenital Heart Disease Program at Nationwide Children's Hospital Heart Center, Columbus, Ohio.

successful, further treatment may not be necessary. Medication can also be used to treat heart rhythm problems. If the problem is life-threatening, a defibrillator might be implanted. A defibrillator is a device that shocks the heart into a regular rhythm.

- **Baffle problems:** Over time the baffles in the Mustard/Senning repair can develop problems. Sometimes they become too narrow (stenotic). ACHD doctors often can treat a narrowed baffle non-surgically. They use a tube into the heart (catheter) to place a wire tube, called a stent, in the narrow area. If a stent cannot fix the problem, surgery may be needed. Baffles can also leak over time. The baffle sutures may come loose and holes can develop. Sometimes these leaks do not need treatment. If they are large, they can cause problems with blood clots and strokes. Doctors often can use a covered stent to stop blood leaks in the baffle. It is important that TGA patients have adult congenital heart disease (ACHD) doctors check their heart regularly to make sure that their baffles still work well.
- **Pump problems:** In a normal heart, the left ventricle pumps blood out to the body. It has thick walls and strong valves designed to do the hard work of pumping to the body. The right ventricle has weaker valves and thinner walls built to pump to the lungs. After a Mustard or Senning repair, the weaker right ventricle pumps the blood to the body. Over time, the right ventricle sometimes has a hard time providing enough pump strength to serve the body. When the heart struggles to keep up with the body's needs, doctors call it heart failure. Heart failure symptoms can include feeling extra-tired on exertion, shortness of breath on exertion, and/or retaining fluid. Heart failure does not mean that the heart stops or fails. Research reports that about 10% of TGA post-Mustard/Senning patients are now experiencing heart failure.

How are pump problems diagnosed?

Echo or MRI can be used to estimate the heart's pumping strength. Because people with TGA post-Mustard have such unusual anatomy, measuring heart function can be challenging. If you have TGA, it is important that your testing be done by an expert in ACHD. Because non-ACHD experts are not trained in CHD hearts, tests done on TGA patients at non-ACHD centers can often be inaccurate.

How are pump problems treated?

The good news is that there are many ways that pump problems in Mustard/Senning patients can be treated and improved. Your ACHD team may recommend exercise to strengthen and improve your heart's pump. There are many medications that also help the heart pump more efficiently. Your ACHD team will also assess whether there are rhythm or baffle problems

that might be making your heart struggle. If these problems can be solved, heart failure may improve.

If other treatment options are not working, surgical options may be considered. In a small number of adult TGA patients, switch conversion might be discussed. This means that the Mustard/Senning repair is taken down and replaced with an arterial switch. There are many challenges to successful switch conversions in adults. It is highly complex, risky surgery. Switch conversions are rarely done in adults. It should only be done at highly-specialized ACHD centers. If heart failure worsens and other treatment options are not available, heart transplantation can be considered.

Is pregnancy safe for TGA post-Mustard/Senning patients?

All women with TGA should consult with an ACHD program before getting pregnant. This will help identify any heart problems that should be addressed before pregnancy occurs. Most women with TGA post-Mustard/Senning can safely undergo pregnancy and delivery. The exceptions are when there is heart failure, problems with lung pressure, and/or uncontrolled rhythm problems. All pregnant women with TGA should be followed by a special maternal fetal health team working together with their ACHD team. In a small number of cases, women with TGA experience new problems with heart failure and/or heart rhythm during pregnancy. These issues may continue even after the pregnancy.

What kind of cardiology care is recommended for adults with TGA post-Mustard/Senning?

The American College of Cardiology and the American Heart Association classify transposition of the great arteries as a highly 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. If you have TGA post-Mustard/Senning, you should see your cardiologist regularly, 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 that can help strengthen and protect your heart work best if started early.

The good news is that most people with TGA post-Mustard/Senning are continuing to do well. By taking good care of your heart and getting recommended care, you can help ensure that you and your heart continue to thrive.

Dr. Gary Webb, who provided his expertise for this handout and accompanying ACHA webinar, directs the ACHD program at Cincinnati Children's Hospital. He previously worked in Toronto, where he directed the world's oldest and largest ACHD program. He has had the pleasure of working with ACHA on many initiatives and served as chair of the Medical Advisory Board from 2006-2011.