Complete Transposition of the Great Arteries
And
The Congenital Heart Surgeon Society.

What is Complete Transposition?

Complete transposition of the great arteries, commonly referred to, as TGA or d-TGA is the most frequent cause of low blood oxygen level in the newborn. These babies have a blue colour, especially of their fingers and toes, a condition referred to as cyanosis. Transposition is a result of abnormal heart development very early during pregnancy. It affects boys more frequently than girls (ratio of 2:1). The cause is unknown and there is no way it can be prevented.

In the normal heart the blue blood returning from veins of the body enters the right side of the heart where it is pumped into the lungs by way of the pulmonary artery. (Figure 1.) Within the lungs, blue blood picks up oxygen (and gets rid of carbon dioxide) and becomes pink. The pink (oxygenated) blood from the lungs then returns to the left side of the heart where it is pumped into the aorta to supply nutrition to the body. Thus there are 2 circulations which are in series: the right-sided blue (pulmonary) circulation and the left-sided pink (systemic) circulation.

Figure 1. Normal Heart

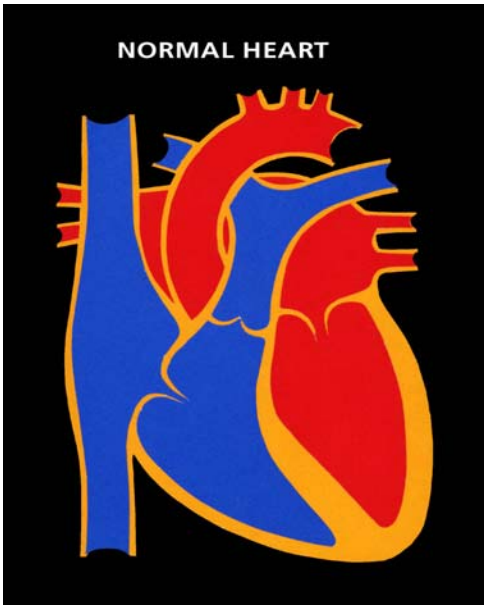
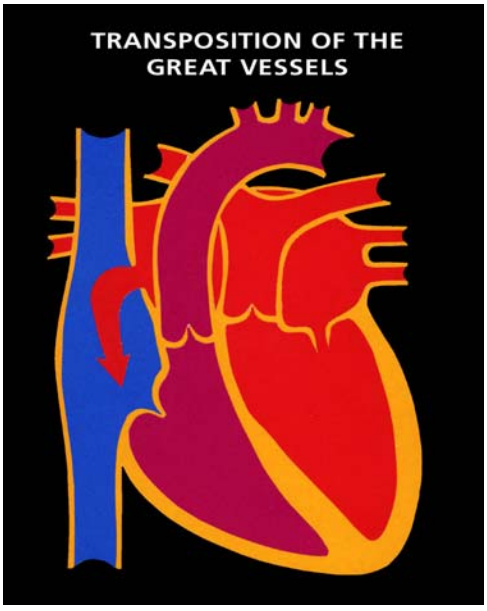


Figure 2. Transposition Heart



In babies with transposition (TGA) the connections of the 2 pumping chambers (the ventricles) to the great arteries (the pulmonary artery and aorta) are reversed, i.e. transposed. (Figure 2) Therefore blue blood returning from the body is pumped back to the body, and pink blood from the lungs is pumped back into the lungs. The 2 circulations are in parallel. If the separation of these 2 parallel circulations is complete

then no oxygen would go to the body and the baby would not survive birth. There is always a hole between the 2 upper chambers of the heart (the atria). This hole, called an atrial septal defect (ASD) is always present before birth. Before birth the ASD in the normal heart allows shunting of blood from the right side of the heart to the left side so that very little blood goes to the lungs that are not breathing air before birth. Following birth, the ASD in babies with TGA is a lifesaver. It allows mixing of the 2 parallel circulations. Mixing of the blue and pink blood allows a limited amount of oxygen to be pumped into the body. The oxygen level in a baby with TGA is well below normal and therefore these babies appear blue. The medical term for a low oxygen level that causes blueness is cyanosis. Without treatment most babies with transposition will die within a few days or weeks of birth. Babies who have an additional hole between the 2 pumping chambers, called a ventricular septal defect (VSD) may survive longer because they have more blood flow into the lungs and therefore better mixing of the 2 circulations. However these babies rapidly develop changes in the lung arteries that are fatal.

([1] See footnote re: Congenitally Corrected Transposition)

Outcomes of Newborns with TGA without Treatment

The outlook for babies with TGA was very poor before surgical treatment was available and their outlook varied according to whether or not they had any additional heart defects. About 75% of all babies with TGA have only an atrial septal defect (ASD) and a patent ductus arteriosus (PDA; an arterial connection between the 2 great arteries, the aorta and the pulmonary artery). Both an ASD and a PDA are present before a baby is born and both normally close by themselves within a few days or weeks of birth. But in TGA, an ASD is essential for survival because it allows mixing between the 2 circulations, as explained above. However, when the atrial septal defect is very small the baby may die within a few minutes after birth. A PDA increases blood flow into the lungs and can cause more mixing of the 2 parallel circulations in TGA, therefore making the baby less blue and their survival slightly better.

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[1] **Congenitally Corrected Transposition** (CC-TGA) is a different form of transposition. As the name implies it is, "congenitally corrected", meaning that at birth the circulation is normal in spite of the heart defect. In this condition the right atrium is connected to the left ventricle that pumps blood into the pulmonary artery. Blood flow returning to the heart from the lungs enters the left atrium and then the right ventricle where it is pumped into the aorta. The connections of atria to ventricles and of ventricles to great arteries are described as discordant, and because both inlet and outlet connections are discordant, the circulation through the heart and lungs is normal; i.e. 2 wrongs make a right! When patients with CC-TGA have no other heart defects, they may be perfectly well for many years. Their heart & circulation are the same as patients with simple TGA after an atrial switch operation.

Almost invariably however, patients with CC-TGA have an associated heart defect. The common defects are a VSD, a leaking inlet valve in the systemic pumping chamber (tricuspid valve), narrowing of the outlet valve in the pulmonary circulation (pulmonary valve), and a block in the heart rhythm that prevents transmission of the electrical impulse between the upper (atria) and lower (ventricles) heart chambers. These 4 common lesions may occur in isolation or in any number of combinations. The presence of the associated heart defects affects the outlook for these patients. Surgery to correct the defects associated with CC-TGA is usually required.

When a ventricular septal defect (VSD) is present (21% of all patients), there is considerably more blood flow to the lungs, so the oxygen level is much better. But in some babies with a VSD there may be too much blood flow to the lungs resulting in flooding of the lungs and heart failure. Babies with TGA and a VSD are less blue (less cyanotic) and about 20% live beyond the 1st year of life but then may die from high blood pressure in the lungs. Others suffer from too much blood flow to the lungs and die within a few months of birth due to heart failure.

The 3rd heart defect associated with TGA is a combination of a VSD and pulmonary valve stenosis (PS). The pulmonary valve allows flow into the lungs while preventing back flow. The combination of TGA with VSD and PS is uncommon, accounting for only 4% of the babies in the Congenital Heart Surgeon Society (CHSS) study. When the amount of pulmonary valve narrowing (PS) is enough to prevent too much blood flow from the VSD into the lungs, the 2 parallel circulations of TGA may be just the right balance to provide enough oxygen without flooding the lungs with too much blood. Survival of these babies before treatment is better than that for babies with simple TGA or TGA and a VSD.

Surgical Treatment of TGA

Until the early 1950's there was no treatment available for babies born with TGA. Drs. Alfred Blalock and Roland Hanlon developed an operation to enlarge the atrial septal defect to improve the mixing of the 2 parallel circulations. Their operation increased the amount of blood oxygen and allowed these babies to survive longer, but the risk of the operation to the baby's life was fairly high and at best, the babies lived only a few months longer. A variant of the "Blalock-Hanlon operation" was devise by Dr. Stirling-Edwards who enlarged the ASD and also changed the flow of blood from the right lung into the right heart by suturing a part of the atrial wall to the left side of the right pulmonary veins. Stirling-Edwards operation was useful in babies who had a VSD needed surgical narrowing (banding) of the pulmonary artery to prevent excess blood flow into the lungs.

During the late 1950's several surgeons attempted to "switch" the 2 great arteries to correct the blood flow. The purpose of the arterial switch operation was to restore the connections of the heart to normal. The arterial switch operation also required transfer of the 2 tiny coronary arteries that are the only source of blood for the heart muscle. If these coronary arteries did not supply enough blood to the heart muscle the babies would die of a heart attack. The primitive medical and surgical techniques of that 1950's era and the lack of intensive care facilities resulted in complete failure.

In 1959 Professor Ake Senning of Zurich devised an "atrial switch" that restored the circulation to normal but left the abnormal connections between the heart and great arteries. He switched the inflow of blood into the heart. In effect his operation created a 2nd. 'Wrong' to make a right! Dr. Senning's operation rearranges the child's atrial wall to make a baffle that redirects the venous blood returning to the heart. It redirects the blue blood from the body into the left side of the heart where it is pumped to the lungs and

diverts the pink blood returning from the lungs into the right heart where it was pumped back to the body. The end result is a normal circulation but with the 2 major pumping chambers (the right and left ventricles) supporting the opposite sides of the circulation.

The Senning operation was technically difficult and other surgeons had difficulty getting good results. In 1963 Dr. William Mustard of Toronto devised a much simpler way of switching the venous inflow but with much lower risk to the child. Mustard created a large atrial septal defect and then inserted a baffle of pericardial (a thin membrane around the outside of the heart). The Mustard baffle switched the flow of blood (i.e. transposed the flow) between the atria and the ventricles. Other surgeons found it easier to do than the Senning operation and it quickly spread around the world.

The atrial switch operation (either Senning or Mustard) dramatically improved the survival of TGA babies from fewer than 20% surviving to their 1st birthday to more than 80% surviving to age 20 years. However, for babies who had an associated VSD the outcome after an atrial switch was less satisfactory.

As experience with the atrial switch operation (both Senning and Mustard) accumulated, two problems became clear. First, an important number of babies born with TGA (about 12%) died before the atrial switch operation could be performed. Second, after many years following a successful atrial switch operation the right ventricle began to fail in some patients. Failure of the right ventricle resulted in a decreasing level of physical activity and often with abnormal heart rhythm. In addition, the outcomes were much worse for babies with TGA and an associated VSD.

In 1978, Dr. A. D. Jatene of Brazil reported success with the arterial switch operation for babies with TGA, the same operation that had failed in the 1950's. Jatene's operation reconnected the pulmonary artery and aorta to the normal position and also transferred the 2 tiny coronary arteries from one great artery to the other. His operation resulted in each ventricle supporting the appropriate circulation; the right side pumping to the low pressure lung circulation and the left to the high pressure systemic (aortic) circulation. Jatene's report, combined with the increasing awareness of the limitations of the atrial switch operation, led to renewed enthusiasm for his more corrective operation called the arterial switch. By that era, considerable progress in cardiology, cardiac intensive care, anaesthesia, cardiac surgery, heart-lung bypass procedures (especially coronary artery surgery), overcame the problems that prevented success in the 1950's. With further experience it became evident that the Jatene operation could be done within a few days of birth thereby preventing deaths that might have occurred if they had to wait for an atrial operation that was typically performed at an older age. With experience, the Jatene operation in babies who had an associated VSD produced results equal to those with isolated TGA.

The Congenital Heart Surgeon Society (CHSS) Study of TGA

In 1985 Drs. John Kirklin and Eugene Blackstone proposed to the members of the CHSS that all babies with complete TGA and under age 2 weeks when admitted to any of the institutions be entered into a study to determine the best management strategy. No attempt was made to influence the management at each institution. When the study began there was uncertainty about which management strategy was best for newborn babies with TGA. The treatment approach used was left entirely to the physicians at each member institution. Between 1985 and 1989, 985 newborn babies TGA were entered into the study. Subsequent analyses of the outcomes led to 9 publications by the CHSS. These can be seen in detail on our website **(www.chssdc.org).**

The initial CHSS publication on TGA with or without a VSD (Trusler et al, 1987) analyzed the 1st 18 months of the study at the original 20 institutions. It demonstrated that both of the atrial switch operations and the arterial switch resulted in similar survival. The data also showed that results for each operation improved during the short period of the 1st 18 months of the study. There was evidence that the survival of babies with TGA and a VSD managed with an arterial switch operation (but not the atrial switch) was as good as that for babies with simple TGA. The early experience demonstrated that the arterial switch operation was as safe as the atrial operation and was advantageous for the more complex babies with TGA and a VSD.

The 2nd publication (Castaneda et al; 1988) examined the outcome for babies with simple TGA (i.e. no associated heart defects other than an ASD or a PDA). Among the first 187 babies admitted into the study with simple TGA, 81% survived to their 1st birthday (recall that before there was treatment for TGA, 80% died before their 1st birthday). The study also showed that survival after the newer arterial switch operation was as good as the atrial switch. The data also predicted that the survival rate with an arterial switch operation could reach 92% at 1 year after hospital admission.

By 1988, the number of babies entered into the study reached 466. A publication by Nowood et al. made 2 important conclusions. It predicted for the first time that the late results for the arterial switch would be better than after the atrial switch. It also showed that for the difficult group of babies with an associated VSD there is an advantage to doing the operation soon after birth rather than the previous policy of delaying surgery to an older age.

Enrolment in the CHSS study was complete by 1989 by which time 985 babies were entered. Among the total there were 514 babies treated with the arterial switch operation. In 1992 Dr. Kirklin and colleagues reported that the arterial switch operation was the best treatment. They looked for anything that might affect outcome with this new operation. The detailed analysis of all 514 “arterial switch” babies identified that certain a normal variations in the pattern of coronary artery anatomy were very important. Some institutions at that time had managed to overcome difficulties with these unusual coronary artery patterns better than others.

One of the CHSS institutions had a unique policy of surgical repair during the newborn period, with either the Senning atrial operation or the arterial switch. Most institutions within the CHSS and elsewhere delayed the atrial switch operation to an older age. Turley et al. in 1995 examined the results of their early repair policy and determined that survival at 6 years was similar with either operation but the arterial switch patients had better heart rhythm and better physical functional ability.

One of the problems of the arterial switch operation was obstruction in the surgical connections between the heart and each great artery. In 1997, Williams et al studied this problem and found that the risk of obstruction on the right side of the heart (0.5% per year) was much higher than on the left side (0.1% per year). Further they identified factors that could be controlled to decrease the risk of developing these complications.

Although the evidence was accumulated that the arterial switch was superior to the atrial operation, there were a considerable number of children successfully treated by the atrial operation. In 2000, Wells et al. examined the late results of all 281 CHSS babies who had an atrial switch between 1985 and 1989. Survival 10 years after operation was 84% and was somewhat better among the Mustard patients (93%) than the Senning (78%). When last seen, most children were either free of limitations (60%) or mildly limited (40%). Serious rhythm disturbance, artificial heart pacemakers, and right heart failure were relatively uncommon.

Quality of life was examined 13 years after the surgical repair of TGA in a study published in 1993 by Culbert et al. A Child Health Questionnaire was completed by 306 children. Their quality of life, as judged by the children themselves, was excellent and comparable to published normal controls. Importantly the study showed that quality of life was better after the arterial switch than after the atrial operation.

In 2003 a CHSS presentation to the European Association of Cardiothoracic Surgery by Williams and colleagues examined the status of all 829 CHSS patients who had undergone a surgical repair of TGA, TGA and VSD, or TGA plus VSD and PS. These children were then 12 to 17 years after their first hospital admission as newborns. Survival at 15 years was 84% and the late risk of death was less after the arterial switch operation. The need for re-operation is low except for those children with pulmonary valve stenosis who require replacement of their artificial pulmonary artery and valve.

Summary

Since the CHSS study began in 1985, the outlook for babies born with TGA has continued to improve. CHSS studies led the world in identifying ways and means to improve care for these babies and children who are now becoming adults. We will continue to follow our patients in an ongoing effort to ensure their well-being.

The amazing improvement in the outlook for newborns with TGA would not have been possible without the co-operation of courageous parents and their children. The CHSS thanks all of those families who have made it possible to help us refine the treatment of babies with TGA and the 'spin-off' effects that have carried over to help children born with other types of heart defects. Together we will continue to make progress. We are very appreciative of your help and thank you all. We hope you will keep in touch with us through our website (www.chssdc.org) or contact us directly at any time.

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