IT BAFFLES ME! A Case Study by Monica McNamara

PREFACE:
Many years ago, I was on my way home from school and I excitedly closed my eyes before pulling onto my street. I knew if my family's car was no longer in the drive way, that meant my mom was at the hospital, giving birth to my younger sister. An enthusiastic, clapping group of children cheered with me in the car as we saw the car was in fact gone, and my mom had gone into labor with my sister. Excitement soon turned into confusion and worry as we learned my sister was born with Transposition of the Great Arteries. Thankfully, my sister's case study is a happy one, but the success and strength of her heart and her quality of life were things she fought for. With the help, talent, skills and love of a loving family, cardiologists, nurses, pediatric cardiologist surgeons, and cardiac sonographers, my sister is still living a healthy, successful life.

This is the story of Erin.

PATIENT HISTORY:

Erin was born cyanotic and short of breath. An echocardiogram shortly after birth revealed that she had dextro-Transposition of the Great Arteries (d-TGA). Erin was not receiving adequate oxygen in her body, which is why d-TGA is one of the heart problems termed, "blue baby syndrome" (stanfordchildrens.org).

Erin's pulmonary and aortic arteries were switched. This caused Erin's oxygen-rich blood to travel to her lungs, via her left ventricle, while oxygen-poor blood circulated through her body, via her right ventricle. Without correction, two separate circuits remain in place for blood flow. One circuit continually circulates deoxygenated blood from the body back to the body. While the other circuit circulates oxygenated blood from the lungs back to the lungs. Without surgery, this disease is fatal.

Cardiologists performed a Balloon Atrial Septostomy when Erin was less than 24 hours old. This procedure would delay surgery until Erin was older. The procedure enlarges the foramen ovale, allowing oxygen-rich and oxygen-poor blood to mix together, enabling oxygen to flow through her body. The Balloon Atrial Septostomy is less invasive than open heart surgery as it is done without general anesthesia and is performed in a cardiac cath lab.

In the case of d-TGA, ventricular and atrial septal defects are beneficial as they help mix the oxygenated blood with the deoxygenated blood, so that babies receive added oxygen.

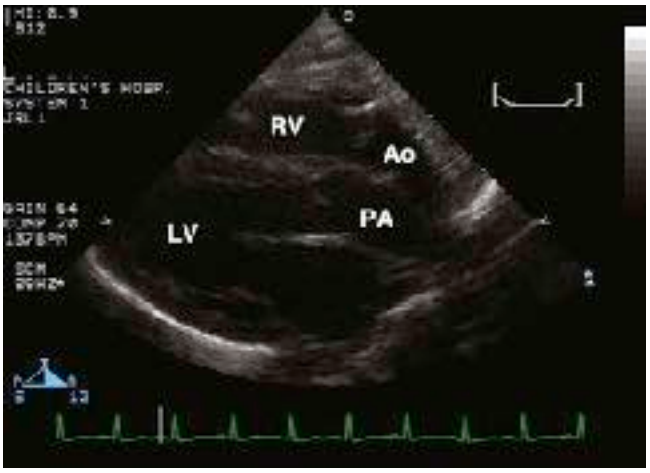
In Erin's case, she had no VSDs and the ASD created by the Balloon Atrial Seprostomy aided her oxygen circulation for about six months. At this time, Erin's body was not getting the oxygen it needed and she would have open heart surgery to re-route the blood in her heart.

Erin's surgeon performed the Senning procedure. This repaired her heart by creating a baffle out of her own tissue in the right atrium. The creation of the baffle in this procedure is incredibly delicate and complex. Some refer to it as, " 'origami-style' cutting and folding of the native atrial tissue to achieve the venous baffle (www.chd-uk.co.uk)."
Erin's pediatric surgeon created two tunnels between the atria. One tunnel reroutes deoxygenated blood coming into the right atrium and carries it to the left ventricle. The other tunnel reroutes oxygen-rich blood from the left atrium, carrying it to the right ventricle. This enables oxygenated blood to travel through her body, however, her right ventricle remains functioning as the systemic pump.

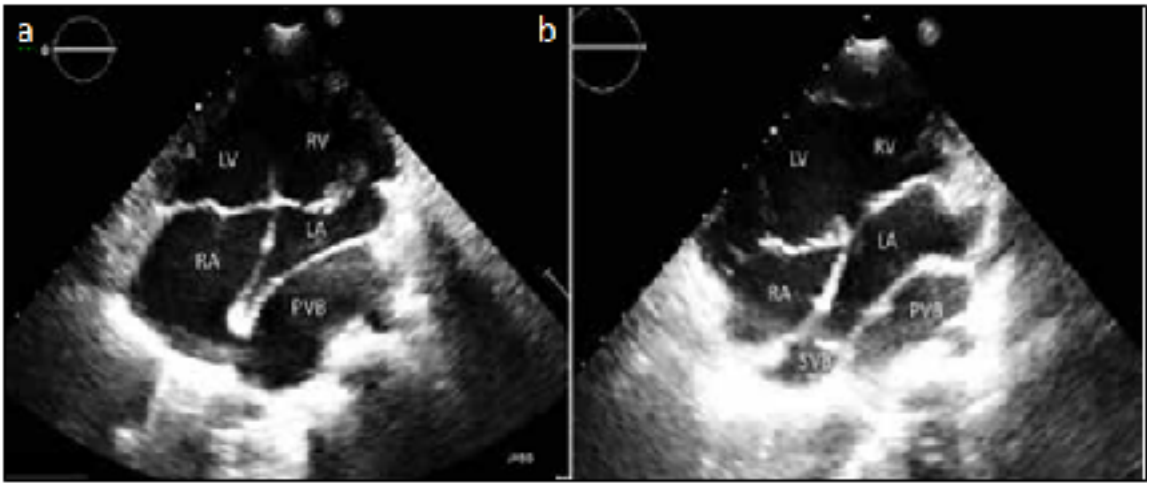
The genius creator of the Senning procedure, along with her very skilled surgeon successfully performed the Senning Procedure on Erin and they saved her life.

Thirty- two strong years later, Erin has had no complications or additional cardiac procedures needed. She is a healthy young woman, a triathlete, and a mother of one, with another baby on the way.

SONOGRAPHIC APPEARANCE:



This is the sonographic appearance of a heart with transposition of the great arteries, before correction.



This is the sonographic appearance after the Senning Procedure.

CURRENT TREATMENT:

Erin receives echoes annually. She also wears a Holter Monitor for 48 hours, to monitor heart activity each year. Each year, her heart showed no further complications that require intervention. Her baffles do show slight leaks, and her EF is 35%. She is continually monitored for deterioration of her heart.

Erin is monitored with extra precaution during pregnancy. Although her cardiologist has seen that pregnancy is well tolerated among women who have had the Senning procedure, Erin is monitored closely as an added safety measure. Blood volume increases during pregnancy by nearly 50%. This puts an added strain on Erin's heart. Erin receives an echo at the beginning of her pregnancies, again at 20 weeks pregnant, and again at 30 weeks. This is to ensure her heart is operating at it's normal capacity, even with the increase in blood volume.

At 36 weeks pregnant, during Erin's first pregnancy, she was symptomatic of heart difficulty. She was dizzy, experienced heart palpitations, chest pains, and light headedness. Her cardiologist wanted to prevent the risk of Erin going into heart failure, so her daughter was born via cesarean birth at Duke Hospital, without complications. Erin spent a night in the cardiac ICU as a precaution, to monitor her heart activity. Her heart showed no signs of further strain and her cardiologist has approved a second pregnancy.

A more modern approach to correcting d-TGA is the arterial switch. During this surgery, the pulmonary artery and the aorta are connected to the appropriate ventricles. The coronary arteries are also attached to the new aorta in their appropriate positions. Any atrial septal defects, or ventricular septal defects are patched, as well as the Patent Ductus Arteriosus, if needed, during this surgery.

The arterial switch has replaced the Senning procedure, unless there are medical reasons to indicate otherwise. The major advantage of the arterial switch is that the right ventricle is able to do the task for which it is created: it pumps blood to the lungs. The left ventricle is the systemic pump after the arterial switch.

In the case of the Senning Procedure, the right ventricle remains the systemic pump, while the left ventricle pumps blood to the lungs. The right ventricle is not created for such a task and in Erin's case, and in cases of all recipients of the Senning Procedure, the right ventricle is over worked and becomes hypertrophied.

PROGNOSIS:

At Erin's most recent echo, her EF was 35%. Her right ventricle is moderately hypertrophied, as it does the work of the systemic pump. Erin's ventricular septum bows into the left ventricle throughout the cardiac cycle. This finding is consistent with a systemic right ventricle. Erin's aorta is mildly dilated at 4.1 cm at the Sinus of Valsalva. Her cardiologist believes, that if circumstances remain the same, Erin will need no other surgical intervention for 15 – 20 years. At this time, she will be a candidate for a heart transplant.

The work that is done in the cardiology world is nothing short of amazing.

REFERENCES:

American Heart Association: heart.org
About Kids Health: aboutkidshealth.ca
US National Library of Medicine: National Institutes of Health: ncbi.nlm.nih.gov
Congenital Heart Defects UK: chd-uk.co.uk
Adult Congenital Heart Association: achaheart.org
Stanford Children's Health: stanfordchildrens.org