Case Number 3
A Univentricular Heart: Tricuspid Atresia

Simon Mifsud and Emma Schembri
Reviewed by: Prof. V. Grech

Case summary:

Demographic details:
Master TA, male, Balluta
Referred from: MDH

A two-year-three-month-old boy was diagnosed with tricuspid atresia. This condition requires three surgical interventions, of which he has already had two. He has now presented to hospital with shortness of breath. This implies that he might now benefit from the third operation. Investigations, which were planned beforehand, will assess whether he is a candidate for this operation, that will improve his symptoms and hence his quality of life.

Presenting complaint:

Shortness of breath
Reduced exercise tolerance

History of presenting complaint:

The shortness of breath has been present for about a year, but has been noticed to have increased progressively during the last three months. Furthermore, during vigorous physical activity, the child ends up vomiting. Nevertheless the child is still very active. There were no other associated symptoms, such as cough, fever, or chest pain.

Past medical and surgical history:

The patient was diagnosed with tricuspid atresia on the third day of life. This was confirmed with an echocardiogram which showed a heart with a missing patent tricuspid valve. This echocardiogram was indicated due to the presence of perioral cyanosis, clubbing of the fingers and toes and a pan-systolic murmur grade 3 and a loud second heart sound (S2). During crying episodes, the patient was noted to become dusky but without any hypercyanotic spells. The oxygen saturations recorded at this point in time were 85% on air.

Two weeks later, the child presented with mild cyanosis and a fever of 38.1^{o}C. Investigations revealed that the oxygen saturations were 78% but all other investigations including the septic screen were negative. As a result the child was administed low flow oxygen, together with ceftriaxone, amoxillin and rocycline.

The dramatic fall in the patient's oxygen saturations initiated a cascade of surgical interventions. The first was the Modified Right-Blalock Interposition Shunt carried out at 24 days of life. This operation involved the temporary insertion of a 3.5mm shunt between the subclavian and pulmonary arteries. Furthermore, the interatrial septum was obliterated by an atrial septectomy. After this intervention, the oxygen saturation improved to 89%.

The second operation was carried out at 9 months of age. This was the Bi-directional Superior Cavopulmonary Anastomosis (BSCA). In this procedure, the previous shunt was removed and the superior vena cava was connected to the pulmonary artery. The saturations improved after this procedure (i.e.: from 74% to 79%) but the increase was not as dramatic as that after the shunt insertion.

Gestational history:

The child was born at 39 weeks of gestation by normal vaginal delivery with a birth weight of 3.47kg. The patient appeared cyanosed at birth due to a slightly traumatic vaginal delivery, but otherwise there were no other significant complications. The Apgar score was 9. The mother had a miscarriage prior to this birth. The parents were not exposed to harmful chemicals during gestation.

Drug history:

Drug	Dosage	Frequency	Type	Reason
Aspirin	50mg	PO, once daily	Anti-platelet agent	Prevents thrombosis

He is also following the national immunisation schedule.
There are no known drug allergies.

Family history:

No family history of congenital cardiac problems.

Social history:

The patient lives at home with his parents and younger sister. He is sociable and plays with peers of the same age.

Systemic inquiry:

- General Health: looks well in general.
- Cardiovascular System: tricuspid atresia
- Respiratory System: shortness of breath, reduced exercise tolerance
- Gastrointestinal System: vomiting during vigorous exercise
- Genitourinary System: nil to note
- Central Nervous System: nil to note
- Musculoskeletal System: nil to note
- Endocrine System: nil to note

Current therapy:

The patient was starved from morning prior to the pre-Total Cavo-Pulmonary Connection (TCPC) diagnostic catheter.

Discussion of results of general and specific examinations:

On general inspection, the patient appeared well. He had a respiratory rate of about 30 breaths per minute, i.e.: slightly tachypnoeic. The pulse rate was 130 beats per minute, which is normal for a child of his age. He had a median sternotomy scar from his previous operations. Clubbing of the fingers and toes was

observed. This is a feature of cyanotic congenital heart disease.
On palpation, the apex beat was not displaced and no heaves or thrills were felt. On auscultation of the chest, a grade 1 pansystolic murmur was heard with radiation to the axilla. The pansystolic murmur may be consistent with mild mitral regurgitation due to mitral annular dilatation. This could be explained by the fact that there might be some volume overload to the left ventricle, therefore resulting in dilatation. Auscultation of the lung fields revealed normal air entry and no inspiratory crackles.

Differential diagnosis:

- Previous corrections of the Tricuspid Atresia (i.e.: BSCA) cannot keep up with the child's demands thus highlighting the need for the third and final operation regarding this condition.
- Anaemia.

Diagnostic procedures:

Laboratory exams:

Test: Pulse Oximetry
Justification for test: To monitor the oxygen saturation of the patient's haemoglobin.
Result: The patient had an oxygen saturation of 76%.
Conclusion: The normal oxygen saturation in a child should be greater than 94%, implying that the patient has low oxygen saturations.

Test: Arterial Blood Gases (ABGs)
Justification for tests: To check for arterial oxygen (and carbon dioxide) partial pressures, pH and base deficit.
Result: Arterial oxygen saturation was 100mmHg (75-100mmHg) and carbon dioxide arterial saturations were 49mmHg (35-45mmHg). pH values were 7.276 (7.35-7.45).
Conclusion: The ABG results are practically within normal limits, when one takes into consideration the child's condition.

Before undergoing the third operation, the patient must satisfy the Fontan's "ten commandments". These ten criteria define the ideal candidate for the operation and should ideally be satisfied, in order to ensure a low level of morbidity and mortality after undergoing the Fontan or TCPC operation. These "ten commandments"/criteria are:

1. Age older than 4 years
2. Sinus rhythm
3. Normal systemic venous return
4. Normal right atrial volume
5. Mean Pulmonary Pressure less than 15mmHg
6. Pulmonary Arteriolar Resistance less than 4 wood units/m^2
7. Pulmonary Artery-Aorta ratio more than 0.75
8. Left Ventricular Function and Ejection Fraction more than 60%
9. Competent Mitral Valve
10. Absence of Pulmonary Artery Distortion

The following instrumental exams were done in order to ensure that the patient satisfies the above "ten commandments".

Instrumental exams:

Test: ECG

Justification for test: Tricuspid Atresia is associated with right atrial volume overload and dilatation, left ventricle hypertrophy and left axis deviation. These three cardiovascular structural changes can be picked up using the ECG.

Result: The ECG confirmed that the patient is in sinus rhythm and none of the above features were recorded.

Conclusion: The patient is in sinus rhythm (Criterion Number 2), with no right atrial dilatation (Criterion Number 4) or left ventricular hypertrophy (Criterion Number 8). The BSCA operation was performed to redirect venous blood from the superior vena cava to the pulmonary artery. In doing so, venous blood from the superior vena cava is bypassing the right atrium and the left atrium and left ventricle, meaning that there is less blood going to these chambers. This helps to prevent the above mentioned changes from taking place inside the heart. It also shows that prior to the operation there were no major dilatations of the right atrium or hypertrophy of the left ventricle.

Test: Echocardiogram

Justification for test: To assess heart movements, heart size and pressure gradients.

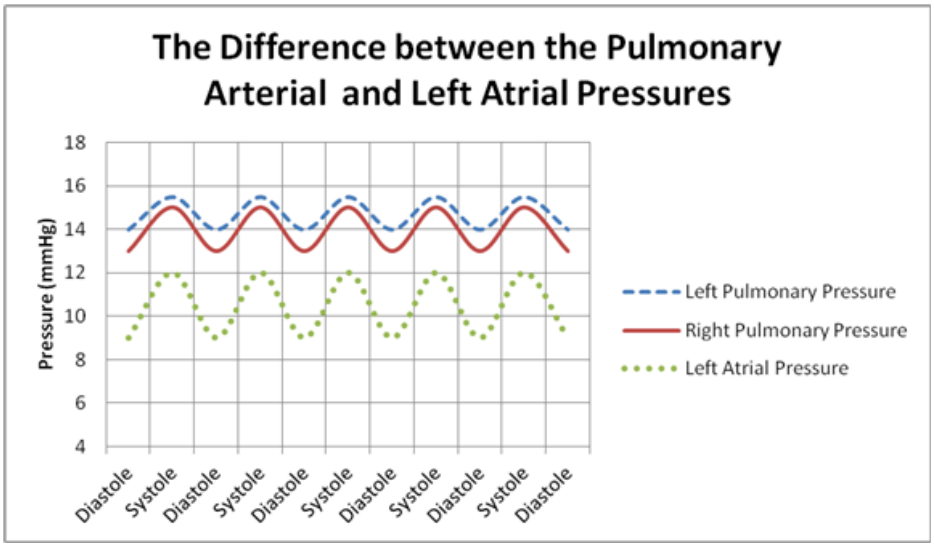
Result: Good Left Ventricular function
Mild mitral regurgitation
Large Atrial Septal Defect
Normal Aorta and Pulmonary arteries

Conclusion: The pansystolic murmur that was referred to the axilla is consistent with mild mitral regurgitation. This does not completely satisfy Criterion Number 9, but if need be, the trivial abnormalities of the mitral valve can be corrected by surgery implying that the patient can still be an ideal candidate for the TCPC Operation. The echocardiogram ensured that the patient has good left ventricular function with an ejection fraction of 86.3% (Criterion Number 8). Moreover, the echocardiogram confirmed the presence of normal aorta and pulmonary arteries (Criteria Numbers 7 and 10). The latter two criteria are not as important as once thought, as nowadays, surgical intervention can correct such abnormalities.

Test: Pre-TCPC Diagnostic Catheter

Justification for test: To assess whether he is a candidate for this operation by measuring the mean pressure gradient between the pulmonary arteries and the left atrium.

Result: The mean pulmonary arterial pressure is 14.38 mmHg (Criterion Number 5) and the mean left atrial/pulmonary venous pressure is 10.5 mmHg. The pressure gradient between the pulmonary arteries and the left atrium is around 4 mmHg (<6mmHg for the TCPC operation to be succesful)[1]. This is depicted in Graph 1. The gap between the pulmonary and left atrial pressures in Graph 1 shows that there is a low pressure gradient between the pulmonary arteries and left atrium. This ensures that blood flows across the pulmonary circulation in order to become oxygenated. If the gap between the pulmonary arterial and left atrial pressures was significant, this would imply that there is high pulmonary vascular resistance. In the Fontan operation, blood flow to the lungs is driven passively without any help from the right ventricle. Thus the presence of a low mean pulmonary arterial pressure (<15mmHg) and a transpulmonary gradient of less than 6mmHg ensures that there is no major pulmonary vascular resistance and that blood flow to the lungs will occur easily.



Graph 1: The Difference between the Pulmonary Arterial and Left Atrial Pressures.

Conclusion: Since the transpulmonary pressure gradient is less than 6, the child will benefit from a TCPC operation, but the operation can be postponed until he is older.

Therapy:

Drugs:

Drug	Dosage	Frequency	Type	Reason
Enalapril	2.5g	Once daily	ACE-inhibitor	To preserve left ventricular function, by reducing afterload and systemic vascular resistance.

Diagnosis:

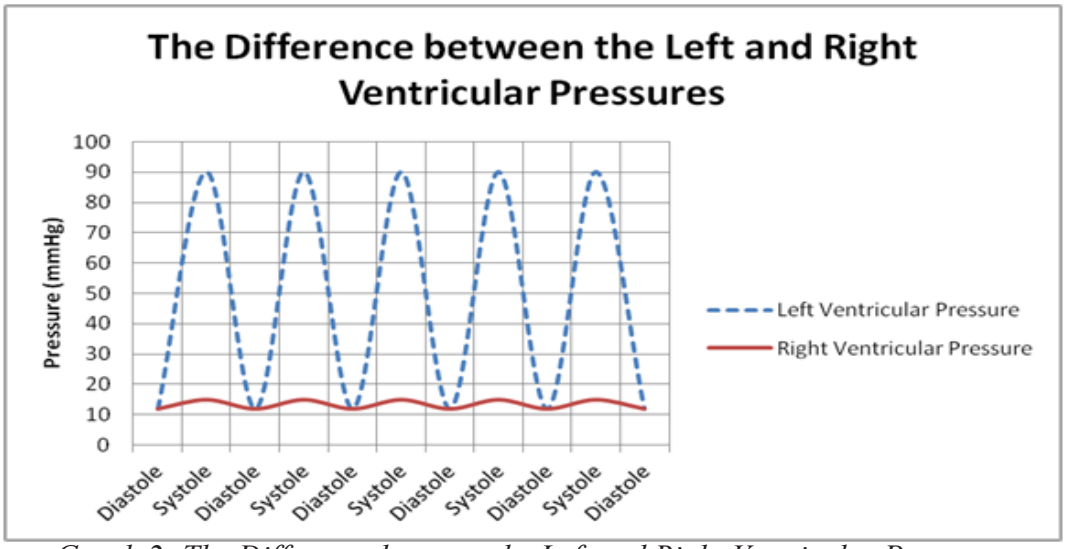
Tricuspid atresia is a type of univentricular heart and is the third most common cyanotic congenital heart disease. There are different subtypes of tricuspid atresia, but this goes beyond the scope of this case report[2]. As the name infers, these patients lack a tricuspid valve between the right atrium and right ventricle[3]. This does not allow blood to flow from the right atrium to the right ventricle and it therefore acts as an obstructive lesion[4].

As a result, the right ventricle does not receive any blood from the right atrium and so remains hypoplastic and undeveloped leaving the left ventricle as the only functional ventricle. Hence the name: univentricular heart. The cause of tricuspid atresia is not known, but it occurs during the first eight weeks of fetal development, when the heart starts to form (i.e.: organogenesis)[3]. Patients with tricuspid atresia depend on an atrial septal defect (ASD) together with a ventricular septal defect (VSD) to maintain an adequate circulation. In fact almost all patients with a tricuspid atresia have these defects i.e.: ASD and a VSD.

Blood reaching the right atrium goes to the left atrium across the ASD. As a result, the deoxygenated blood from the right atrium mixes with the oxygenated blood from the pulmonary veins inside the left atrium. This mixing explains the cyanosis associated with tricuspid atresia. This mixed blood then passes into the left ventricle. Once inside the left ventricle, most of the blood passes into the aorta during left ventricular systole, but some blood also passes through the VSD into the hypoplastic right ventricle and eventually into the pulmonary arteries.

Furthermore, some blood from the aorta flows through the patent ductus arteriosus (PDA) into the pulmonary arteries thus allowing more blood to reach the lungs. A PDA initially allows the patient to live with minimal cyanosis. However problems begin, when the ductus arteriosus closes and since the VSD tends to be restrictive, blood flow to the pulmonary arteries is reduced with a resultant increase in cyanosis. If the blood supply to the lungs is highly restricted, the patient might collapse suddenly. This patient lacked a PDA and as a result presented with perioral cyanosis after birth.

The pan-systolic murmur (referred to as a flow murmur) that was heard soon after birth occurs due to the restrictive VSD. Since the left ventricle is well developed and more muscular than the right ventricle, it generates a larger amount of pressure i.e.: circa 90 mmHg during systole. The hypoplastic right ventricle generates very little pressure during systole i.e.: circa 15 mmHg. During diastole the pressures generated by the right and left ventricles are very similar. Therefore it is only during systole that a large pressure gradient exists between the two ventricles. Therefore left-to-right shunting of blood occurs mostly during systole, causing a pan-systolic murmur to be heard. This principle is depicted in Graph 2.



Graph 2: The Difference between the Left and Right Ventricular Pressures

Final treatment and follow ups:

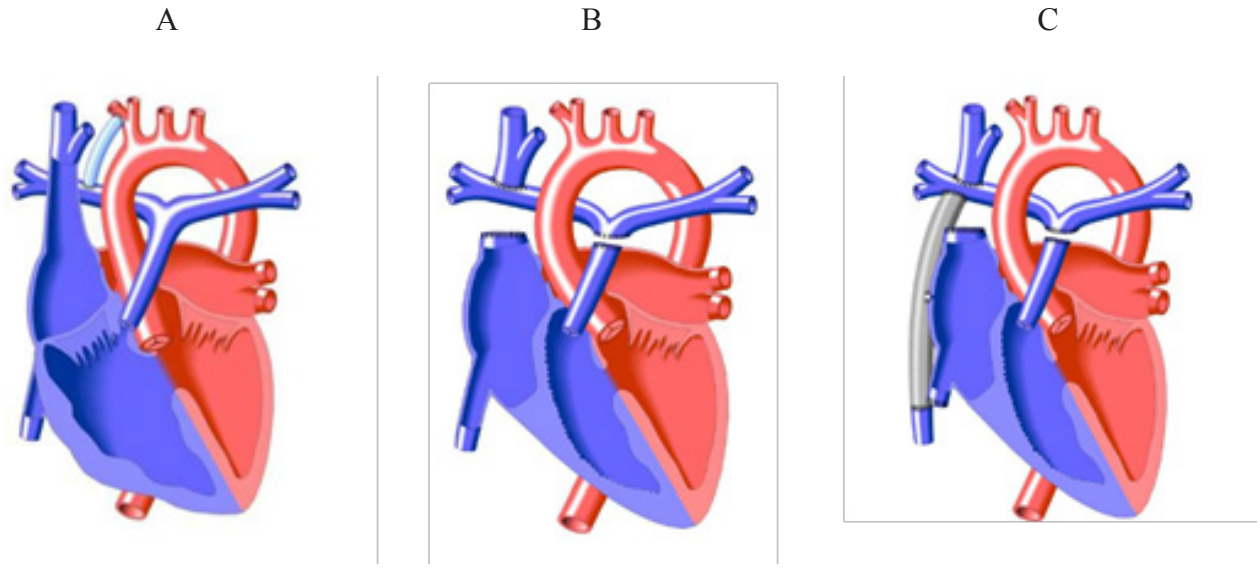


Figure 1: Surgical Interventions in Patients with Tricuspid Atresia. The above figure is a summary of the three surgical opererations required for palliation of Tricuspid Atresia[5]. Key to Figure 1:
A. Modified Right Blalock Interpositional Shunt (Right Subclavian Artery to Right Pulmonary Artery)
B. SCA (Superior Vena Cava to Right branch of Pulmonary Artery)
C. TCPC (Inferior Vena Cava to Right branch of Pulmonary Artery)

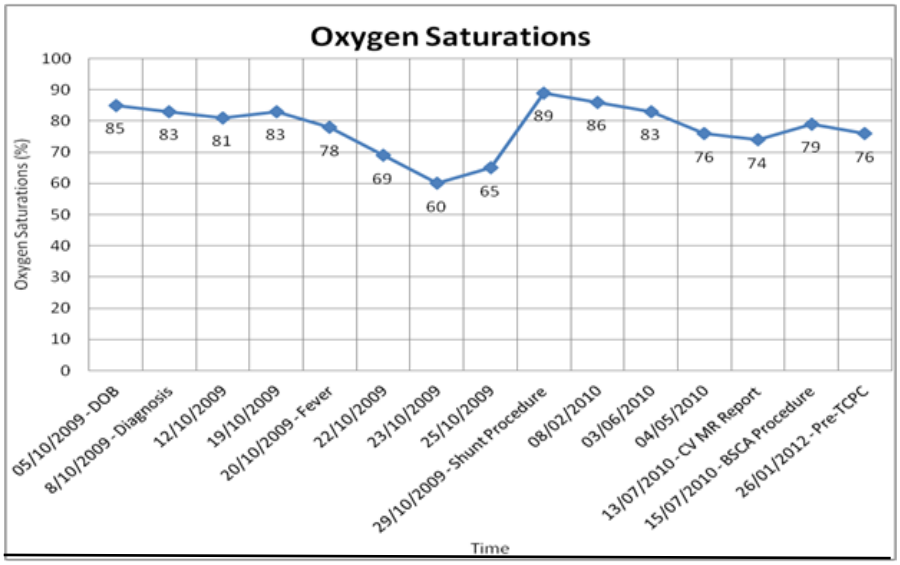
The first operation for tricuspid atresia was done to adjust pulmonary blood flow. Since this child lacked a PDA and appeared cyanosed at birth, his pulmonary blood flow needed to be increased. This increase in pulmonary blood flow was achieved by the insertion of a shunt between the right subclavian artery and the right pulmonary artery i.e.: Modified Right Blalock Interpositional Shunt. This shunt was a temporary one, until the second operation took place. The shunt mimics the function of a PDA, as it allows blood from the aorta via the right subclavian artery to flow into the right pulmonary artery. The shunt tries to compensate for the reduction in the left-to-right shunt across the restrictive VSD.

During this operation, an atrial septectomy was also done. This involved inserting a balloon catheter to enlarge the ASD. The septectomy helps to increase the right-to-left shunt across the atria, in order to prevent right atrial dilatation and allow more blood to reach the left ventricle. This means that the left ventricle has a greater end-diastolic volume and according to the Frank-Starling Relationship, there is a greater stroke volume. This increase in stroke volume is important to make up for the blood that is shunted across the Blalock-Interposition Shunt. The aim of this procedure was to increase the saturations and in fact these increased to 89% at rest, from the previous 65% prior to the operation.

The second (BSCA) and third (TCPC) operations are done separately, rather than together, to reduce morbidity and mortality[1].

The second operation, known as BSCA (or Bi-directional Glenn or Hemi-Fontan), involves connecting the superior vena cava (SVC) to the right branch of the pulmonary artery and removing the Blalock-Taussig Interposition Shunt since it's no longer required. The scope of the BSCA is to shift the blood supply to the pulmonary circulation from one of a high pressure (from the left ventricle) to one of a low pressure (directly from the superior vena cava).

With the Blalock-Taussig Interposition Shunt, the pulmonary circulation is receiving high pressurised blood from the left ventricle meaning that the pulmonary arteries may become thick walled and hard. These changes may result in an increased resistance to blood flow. If this is left for a significant period of time, the resistance offered by the pulmonary circulation, might not allow the patient to be an ideal candidate for the BSCA or TCPC Operations.



Graph 3: Oxygen Saturations. This shows how the oxygen saturations varied during different life events of this patient. Key to Graph 3: DOB – date of birth, CV MR – Cardiovascular Magnetic Resonace, BSCA – Bidirectional Superior Cavopulmonary Anastomosis, TCPC – Total Cavopulmonary Connection.

In the BSCA, the SVC is detached from the right atrium and attached to the right branch of the pulmonary artery. In this way, venous blood directly from the SVC will flow passively into the pulmonary arteries. Without any pump, blood flow across the pulmonary circulation is driven by the central venous pressure and enhanced by the negative intrathoracic pressures that occur during inspiration. As a result this system necessitates a low pulmonary vascular resistance. Further stressing this fact is the age at which this operation is carried out i.e.: at 6 - 9 months. This is the optimum age as the pulmonary vascular resistance would have decreased considerably when compared to that of birth. In this case, the operation was performed at nine months of age. The saturations increased to 79% on air from 74%.

Graph 3 depicts how the onset of fever caused the patient's oxygen saturation to drop dramatically and initiate a cascade of surgical interventions. Overall, after every intervention, the patient's oxygen saturations improved (especially after the insertion of the Blalock-Taussig Interposition Shunt), but with the child growing and becoming more active, the intervention's benefits become limited due to the continuosly increasing demands made by the patient's active body. This explains the gradual drop of oxygen saturations that occur after every intervention.

The third and final operation is known as the TCPC or Fontan procedure. Prior to this operation, a pre-operative assessment is performed with a diagnostic cardiac catheter. The main aim of this test is to establish whether the patient will be likely to benefit from the TCPC operation on the basis of a transpulmonary gradient which is less than 6mmHg. As discussed earlier the results from the pre-TCPC diagnostic catheter show that the child is a candidate for the Fontan operation[1].

After this assessment the child was prescribed enalapril, an Angiotensin Converting Enzyme (ACE) inhibitor, to be taken until the TCPC is done. This was prescribed in order to preserve left ventricular function, by decreasing both afterload and systemic vascular resistance. In doing so, the left ventricle does not have to generate high pressures to eject blood during systole, thus reducing the risk of left ventricular hypertrophy.

The TCPC is planned to take place when the child is about four years of age. In the TCPC the inferior vena cava (IVC) will be connected to the right branch of the pulmonary artery. In the end the right branch of the pulmonary artery will end up attached to the SVC and IVC. The TCPC reduces the volume overload presented to the left ventricle, improves oxygen saturations as it is arranged in such a way that there is no longer any mixing of blood and it finally attaches the low pressure pulmonary system with a low pressure systemic venous system.

This child's mother has recently set up an assocation, known as Beating Hearts Malta. This will help to increase the awareness and show support to patients living with a congenital heart defect. More information about this association can be found on their Facebook Page - Beating Hearts Malta.

Figure 2: The logo of Beating Hearts Malta.

Fact Box 3:

Name of Condition: Tricuspid Atresia

Tricuspid atresia is the third most common cyanotic congenital heart disease. Patients with tricuspid atresia lack a tricuspid valve between the right atrium and right ventricle. Therefore, there is no right atrioventricular communication and as a result the right ventricle remains hypoplastic and undeveloped. These patients are thus left with only one functioning ventricle i.e.: the left ventricle, hence the term univentricular heart. Other defects that are present in patients with tricuspid atresia include an atrial septal defect (ASD), a ventricular septal defect (VSD) and possibly a patent ductus arteriosus (PDA).

Risk Factors: The exact aetiology of tricuspid atresia is unknown, however there are a number of risk factors that may increase the chances of a person developing this condition. These include:

- A Family History of Congenital Heart Defects
- Antenatal problems such as an infection with the Rubella virus, uncontrolled diabetes mellitus during pregnancy and alcohol consumption during pregnancy.
- The use of certain teratogenic drugs during pregnancy such as Lithium, Sodium Valproate, Carbamezapine and other anticonvulsants.
- The presence of Trisomy 13, 18 and 21 is associated with a greater likelihood of the presence of congenital heart defects.

Clinical Features: At birth the patient may be well or have some minimal cyanosis as there is the PDA which augments pulmonary blood flow. However the PDA usually closes 24 to 48 hours after birth and when it closes, there is reduced blood flow to the lungs, and the patient may become severely cyanosed and breathless and may also collapse.

Symptoms of Tricuspid atresia include:

- Breathlessness
- Fatigue
- Poor exercise tolerance and weakness
- Poor Feeding

Signs of Tricuspid Atresia include:

- Cyanosis
- A Pansystolic murmur
- Tachypnoea
- Poor weight gain and slow growth

Diagnosis and Management: The diagnosis of tricuspid atresia is confirmed by an echocardiogram. Once the diagnosis of tricuspid atresia is established, one has to ensure that there is adequate pulmonary blood flow. If there is a PDA, one can start the patient on a prostaglandin E1 infusion to maintain the ductus arteriosus patent. On the other hand, if there is no PDA, the surgical insertion of a left-to-right shunt is required. The latter procedure is known as a Modified Right Blalock Interpositional Shunt. This involves the insertion of a shunt between the right subclavian artery and the right pulmonary artery branch. This operation is usually carried out within the first few days of life.

The second operation occurs when the patient is 4 to 9 months old and involves the removal of the previous Blalock shunt and the connection between the superior vena cava and the right branch of the pulmonary artery. This is known as a Bi-directional Superior Cavopulmonary Anastomosis (BSCA). The

third and final operation is the Total Cavopulmonary Connection (TCPC) and involves the connection of the inferior vena cava and the right branch of the pulmonary artery. This is normally performed when the patient is 1 to 4 years of age. The BSCA and TCPC operations help to improve the patient's oxygen saturations, relieve the patient from cyanosis, and reduce the volume overload presented to the left ventricle.

References:

1. DeGiovanni JV, Grech V. Cardiac catheter assessment of congenital heart disease prior to total cavopulmonary connection. Images Paediatr Cardiol. 2005; 7(4): 10-27.
2. http://emedicine.medscape.com/article/900832-overview - accessed on 16th Dec 2012
3. http://www.lpch.org/DiseaseHealthInfo/HealthLibrary/cardiac/ta.html - accessed on 16th Dec 2012
4. Attard-Montalto S. and Saha V. Master Medicine Paediatrics. 2nd ed. Philadelphia: Elsevier, 2006.
5. http://www.pcics.org/pdf/Tricuspid%20Atresia.pdf – accessed on 15th Dec 2012