It’s Complicated…

Aimee D. Veridiano, MD
Cardiology Fellow

ST. LUKE’S HEART INSTITUTE
DEPARTMENT OF ECHOCARDIOGRAPHY
Objectives

• To present a case of a 30 y/o male with a complex congenital heart disease
• To review the echocardiographic features of Tricuspid Atresia
• To discuss the ACC/AHA guidelines in management of Tricuspid Atresia
General Data

- J.C.P
- 30y/o
- Male
- Married
- Catholic
- Residing in Q.C.
Chief Complaint

- exertional dyspnea
Clinical History

At birth
- (+) cyanosis
- Diagnosed with CHD

Childhood & Adolescence
- active
- At par with age

At present
- No major hospitalizations
- With occasional effort-related dyspnea
- Otherwise unremarkable
Childhood Developmental History

- Delivered term via NSD
- 2nd of 5 children with 3 sisters and 1 brother
- Complete immunization history
- Engaged in strenuous physical activities (basketball weekly) during childhood and adolescence
- Developmental milestones were at par with age
Maternal History

• unremarkable
Past Medical History

- Non-hypertensive, non-diabetic
- No known Allergies
Family History

- (+) Family history of heart disease
- cousin with TOF in the maternal side and a cousin in the paternal side who has an ASD and underwent repair
- cancer & DM = mother side
- hypertension & asthma = father side
Personal/Social History

• Nonsmoker
• Occasional alcoholic beverage drinker
• College graduate
• Works in the accounting department
Review of Systems

- **General**: no weight loss, no anorexia, no fever
- **Skin**: No rashes, no pigmentation
- **HEENT**: No headaches, no visual disturbances, no dysphagia
- **Respiratory**: no cough, no hemoptysis
- **Gastrointestinal**: no melena, no hematochezia, no diarrhea
- **Genitourinary**: No dysuria, no oliguria
- **Muskuloskeletal**: No joint pains, no edema
- **Endocrine**: No polydipsia, no polyuria, no polyphagia, no heat intolerance
Physical Examination

• General appearance: Conscious, coherent, ambulatory

• Vital Signs:
  BP: 110/60 mmHg  HR: 70 bpm
  RR 19 cpm  Temp: 37°C

• Weight: 63.5 kg  Height: 1.65 m  BMI: 23.32 kg/m²
Physical Examination

- **SKIN**: Warm to touch, no skin lesion, and good skin turgor
- **HEET**: Pink palpebral conjunctivae, anicteric sclerae, no naso-aural discharge, non hyperemic tonsils
- **NECK**: No neck vein engorgement, JVP at 4cm, no lymphadenopathy
- **CHEST and LUNG**: Symmetrical chest expansion, no retractions, clear breath sounds
Physical Examination

- **HEART:**

  Apex beat at 6th left intercostal space anterior axillary line, (+) LV heave

  Normal rate, regular rhythm, fixed splitting of S2, loud P2, (+) grade 2/6 systolic murmur at the 2nd ICS left parasternal border
Physical Examination

• **ABDOMEN:** flat, no superficial veins, no pulsating mass, no bruit, normo-active bowel sounds, soft, no tenderness, no hepatosplenomegaly

• **EXTREMITIES:** No gross deformities, (+) clubbing of nails, (+) cyanosis, no edema, pulses are full and equal
Diagnostics

**ECG**
- ECG: NSR, Left axis deviation, occasional PVCs, non-specific T wave changes

**CXR**
- CXR: no focal infiltrates in both lungs, cardiomegaly

**Blood**
- CBC: Hgb 21 g/dl, Hct 65%, RBC 8.13 mil/cumm, WBC 5350, Neutro 53% Lympho 38% Eos 1% Mono 8%, Platelet count 193,000
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-ECHO WITH DOPPLER
2D-Echo with Doppler
2D-ECHO WITH DOPPLER
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Diagnosis

- Congenital Heart Disease
- Tricuspid atresia
- Atrial Septal Defect, secundum
- Ventricular Septal Defect, perimembranous
TRICUSPID ATRESIA

- accounts for 1-3% of all CHD
- Incidence: 0.06 per 1000 live births
- Sporadic, familial, have 22q11 microdeletions
- tricuspid valve is absent, and there is no anatomic connection and direct communication between the RA & RV
- a fibrotic septum is usually present at the site of the tricuspid valve
Normal heart:
- Right atrium
- Tricuspid valve
- Right ventricle

Tricuspid atresia:
- Atrial septal defect
- Tricuspid valve (not developed)
- Ventricular septal defect
Tricuspid Atresia

- AO = Aorta
- PA = Pulmonary Artery
- LA = Left Atrium
- RA = Right Atrium
- LV = Left Ventricle
- RV = Right Ventricle

- Opening Between Atria
- Closed Tricuspid Valve
- Underdeveloped Right Ventricle

- Oxygen-rich Blood
- Oxygen-poor Blood
- Mixed Blood
Systemic venous blood is received by the right atrium

Passes through an interatrial communication to converge with pulmonary venous blood in the morphologic left atrium

Mixed blood flows across a morphologic mitral valve into a morphologic left ventricle

The LV acts as the only pumping chamber for the systemic and pulmonary circulation
TRICUSPID ATRESIA

- In this defect, systemic venous blood runs from the RA, via the PFO or secundum ASD, to the left atrium, where the blood blends with oxygenated blood from the pulmonary veins.
- The only functional ventricular chamber in this heart is the left ventricle.
- RV is rudimentary, there is no inlet and trabecular part, the infundibulum communicates with the LV via the bulboventricular orifice.
TRICUSPID ATRESIA

- Pulmonary artery arises from the hypoplastic right ventricle
- In 90% of patients, the great arteries are in a normal position and the bulboventricular orifice is restrictive (subpulmonary stenosis with protection of the pulmonary vascular bed against pulmonary vascular disease)
- The great arteries are transposed in 10% of the patients with tricuspid atresia, and the bulboventricular orifice is nonrestrictive
• Complications due to chronic hypoxemia; risks in the long run include progression of mitral regurgitation
• Failure of the left ventricle with volume overload
### Classification of Tricuspid Atresia Based on Relationship of Great Arteries & Pulm. Blood Flow

<table>
<thead>
<tr>
<th>Type</th>
<th>Relationship</th>
<th>Frequency</th>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>Normal Great Arteries</td>
<td>69%</td>
</tr>
<tr>
<td>IA</td>
<td>No VSD, Pulmonary Atresia</td>
<td>9%</td>
</tr>
<tr>
<td>IB</td>
<td>Restrictive VSD, PS</td>
<td>51%</td>
</tr>
<tr>
<td>IC</td>
<td>Non-restrictive VSD, no PS</td>
<td>9%</td>
</tr>
<tr>
<td>II</td>
<td>D-TGA</td>
<td>28%</td>
</tr>
<tr>
<td>IIA</td>
<td>VSD, PA</td>
<td>2%</td>
</tr>
<tr>
<td>IIB</td>
<td>VSD, PS</td>
<td>8%</td>
</tr>
<tr>
<td>IIC</td>
<td>VSD, No PS</td>
<td>18%</td>
</tr>
<tr>
<td>III</td>
<td>L-TGA</td>
<td>3%</td>
</tr>
</tbody>
</table>
Echocardiography

- Establishes diagnosis of tricuspid atresia
- Establishes the position of the great arteries, ventricular septum and the nature of the interatrial communication
- Provides important information of the VSD
- Useful in determining the size of the pulmonary trunk and aortic root and in identifying two functional semilunar valves
- Establishes size of LV, LV function and abnormalities of mitral valve
Echocardiography

- An echo dense shelf is in the position normally occupied by the tricuspid valve
- Normally related great arteries
  - RV cavity is small
  - Restrictive perimembranous VSD
  - LV is enlarged
  - RA is increased in size if ASD is restrictive
Echocardiography

- Color flow mapping with spectral Doppler
- Confirms presence and establishes size of the interatrial communication and size of VSD
- Determines presence and degree of MR
Consistent features

- Physiologic and gross morphologic absence of a connection between the morphologic right atrium and morphologic right ventricle
- Varying degrees of hypoplasia of the morphologic right ventricle
- An interatrial communication
- Normally formed morphologic left ventricle with a morphologic mitral valve
Variable features

- The ventricular origin of the aortic root and pulmonary trunk (normally related great arteries or complete transposition)
- The absence, presence and degree of obstruction to pulmonary blood flow
Pathogenetic mechanism

• During early embryogenesis, the inlet component of the developing RV is extremely small and normally expands considerably.
• The process of inlet expansion coincides with the development of atrioventricular valves.
• Failure of expansion of the inlet component of the developing RV and the accompanying tricuspid valve.
Unoperated Survival
The unoperated 10-year survival rate in patients with tricuspid atresia is 46%, with deaths due to hypoxia, cardiac failure, endocarditis, paradoxical emboli, and cerebral abscess. Long-term unoperated survival depends on adequate but not excessive pulmonary blood flow. Such a balanced circulation is rare but occasionally allows unoperated survival into the sixth decade of life.

Operations
All surgical approaches are staged and palliative, because a biventricular repair is not possible.

Current management strategies aim for a Fontan-type circulation in all patients with tricuspid atresia (see Chapter 11). The Fontan operation is performed in hearts with a univentricular atroventricular connection to abolish cyanosis. The ventricular mass is used to support the systemic circulation by excluding a right-sided “pump” from the circulation. Thus, systemic venous blood is directed straight into the pulmonary artery via the right atrium (Fontan operation) or via an intracardiac or extracardiac conduit (total cavopulmonary connection [TCPC]).

Where there is severe pulmonary stenosis the aim of the initial operation is to improve pulmonary blood flow. If intervention is needed in the neonatal period, before pulmonary vascular resistance has fallen,
UNOPERATED OR POST-SHUNT PATIENTS WITH TRICUSPID ATRESIA

History should seek evidence of complications of cyanosis and exercise capacity. Examination findings, including oxygen saturation, should be compared with values obtained at previous visits.

All patients should have periodic assessment with:
- Electrocardiography—to check for sinus rhythm, atrial arrhythmia
- Chest radiography—to evaluate cardiothoracic ratio, pulmonary vasculature
- Echocardiography—to assess ventricular cavity size and function, atrioventricular and aortic valve regurgitation, and patency of shunts
- Exercise testing—to assess functional capacity and degree of desaturation on exercise
- Ambulatory electrocardiographic monitoring—if the patient describes palpitations or faintness
- Thyroid function—for patients on amiodarone. The incidence of amiodarone-associated thyroid dysfunction is high in cyanotic patients; thyrotoxicosis can cause worsening atrial arrhythmia and heart failure.4

All unoperated or shunted patients are cyanosed and have restricted physical activity. It is not usually necessary to impose additional limitations on physical activities because most patients will have adapted their lifestyle so that they live within their limits.
Survival

- Depends chiefly upon the regulation of pulmonary arterial blood flow
- Anatomic arrangements beyond the mitral valve
- Adequacy of the interatrial communication
Survival

• Overall survival
  – 72% at one year
  – 52% at 5 years
  – 46% at 10 years

• Shortest lifespan
  – <6 months
  – Normally related great arteries with intact ventricular septum
Survival

• Shortest lifespan
  – <6 months
  – Normally related great arteries with intact ventricular septum

• Exceptional survival
  – 65 years old
  – Tricuspid atresia, pulmonary atresia, ostium secundum ASD, large aortic to pulmonary arterial collaterals
AHA/ACC 2008 Guidelines for the Management of Adults with Congenital Heart Disease
Table 3. Types of Adult Congenital Heart Disease of Great Complexity*

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Conduits, valved or nonvalved</td>
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<tr>
<td>Cyanotic congenital heart (all forms)</td>
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<tr>
<td>Double-outlet ventricle</td>
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<tr>
<td>Eisenmenger syndrome</td>
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<tr>
<td>Fontan procedure</td>
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<tr>
<td>Mitral atresia</td>
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<tr>
<td>Single ventricle (also called double inlet or outlet, common, or primitive)</td>
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<tr>
<td>Pulmonary atresia (all forms)</td>
</tr>
<tr>
<td>Pulmonary vascular obstructive disease</td>
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<tr>
<td>Transposition of great arteries</td>
</tr>
<tr>
<td>Unbalanced atrioventricular septal defect</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
</tr>
<tr>
<td>Other abnormalities of atrioventricular or ventriculoarterial connection not included above (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)</td>
</tr>
</tbody>
</table>

*These patients should be seen regularly at adult congenital heart disease centers.

1.10.1. Recommendations for Hematologic Problems

Class I
1. Indications for therapeutic phlebotomy are hemoglobin greater than 20 g per dL and hematocrit greater than 65%, associated with headache, increasing fatigue, or other symptoms of hyperviscosity in the absence of dehydration or anemia. (Level of Evidence: C)

Class III
1. Repeated routine phlebotomies are not recommended because of the risk of iron depletion, decreased oxygen-carrying capacity, and stroke. (Level of Evidence: C)
14.3. Recommendation for Surgical Options for Patients With Single Ventricle

Class I
1. Surgeons with training and expertise in CHD should perform operations for single-ventricle anatomy or physiology. (Level of Evidence: C)

14.7.1. Recommendations for Medical Therapy

Class I
1. Warfarin should be given for patients who have a documented atrial shunt, atrial thrombus, atrial arrhythmias, or a thromboembolic event. (Level of Evidence: C)

Class IIa
1. It is reasonable to treat SV dysfunction with ACE inhibitors and diuretics. (Level of Evidence: C)

14.4. Recommendation for Evaluation and Follow-Up After Fontan Procedure

Class I
1. Lifelong follow-up is recommended for patients after a Fontan-type operation; this should include a yearly evaluation by a cardiologist with expertise in the care of ACHD patients. (Level of Evidence: C)

14.5. Recommendation for Imaging

Class I
1. All patients with prior Fontan-type repair should have periodic echocardiographic and/or magnetic resonance examinations performed by staff with expertise in ACHD. (Level of Evidence: C)
Take home points

- History and Physical Exam
- Established by 2D Echo with Doppler studies
- Morphology of structures
- Patient-directed management/therapy
THANK YOU!!!
Perinatal Circulation

Oxygen saturation of blood:
- Red: High oxygen content
- Dark Purple: Medium oxygen content
- Blue: Poor oxygen content

From Moore & Persaud 1998
Summary

- Tricuspid atresia with normally related great arteries and a restrictive VSD
- Cyanosis
- Normal or decreased pulmonary arterial blood flow
- Dominant LV