Cyanotic Diseases
Congenital Heart Disease, Teaching Course
EUROECHO 2010

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Cyanotic Congenital Heart Disease

Classification

- Cyanosis with low pulm. flow
  - TOF
  - Ebstein Anomaly
  - Tricuspid Atresia
  - Pulm. Atresia

- Cyanosis with high pulm. Flow
  - TGA
  - Double outlet V
  - Double inlet V
  - TAPVD
Tetralogy of Fallot

- Etienne- Louis Arthur Fallot made the first published bedside diagnosis that was proven at post-mortem in 1888 and called the condition “maladie bleue”
Epidemiology

- Prevalence varies from 0.26- 0.48/ 1000
- Incidence: 3/10000 live births
- Most common cyanotic CHD
- 6.8 of all CHD
- 10 % of all GUCH patients
Morphology- Ventricular Septal Defect

- 80% of cases fibrous continuation between the mitral, tricuspid, and aortic valves
- 20% cases there is a muscular rim around the defect.
- The pulmonary valve annulus tends to be hypoplastic or atretic.
Morphology

- **Overriding Aorta**
  Degree of override can vary from an exclusive connection of the right ventricle to an exclusive connection to the left ventricle.

- **Pulmonary Stenosis**
  Infundibular / valvular

- **Concentric RV Hypertrophy**
  Secondary to RV outflow obstruction
Associated Lesions

- Pulmonary valvar stenosis
- Pulmonary atresia
- Absence of pulmonary valve leaflets
- Right Aortic Arch (25 %)
- PFO or ASD (Pentalogy of Fallot)
- Anomalous origin of LAD from RCA (3%)
Surgery

- Palliative procedures to increase pulmonary blood flow
  - Blalock Tausing shunt: L Sub CA to PA
  - Waterston shunt: Asc Ao to RPA
  - Potts shunt: Desc Ao to LPA

- Repair
Surgical Repair

- Closure of VSD
- Relieving of RVOT
  - Pulmonary valvotomy
  - Resection of infundibular muscle
  - RVOT or subannular patch
  - Transannular patch - if PV annulus restrictive
  - Pulmonary valve implantation
  - Extracardiac conduit between the RV and PA
  - Closure of PFO or ASD
Survival Following Complete Repair of TOF

- Control Population
- Sudden Death accounted for 70%
- Older age > 12y is a strong predictor
- Transannular patch associated
- n=163
- p<0.01
Follow-Up of Repaired ToF

- Significant PR - Following transannular patch
- Residual RVOT obst.
- RV Dilatation, TR, RV Failure
- Residual VSD
- Progressive AI with or without root dilation
- LV Dysfunction
- Infective Endocarditis
- Electrical Complications: RBBB, Bifascicular block, 3° AV block, A Fl., AF, NSVT, VT
- Sudden Cardiac Death

www.escardio.org/EAE
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Ebstein’s Anomaly

- Apical displacement of the septal and posterolateral leaflets of the TV
- Atrialization of RV inflow - smaller “functional” RV
- Tricuspid regurgitation (occasionally stenotic)
- Right Atrial enlargement
- PFO or ASD (50%)
- Accessory pathways - risk of atrial tachycardias
Ebstein’s Anomaly

May be Associated with

- PFO
- ASD
- VSD with or without Pul. atresia
- Pulmonary Out Flow Obstr.
- PDA
- Ao Coarctation
- LV Cardiomyopathy
**Ebstein Anomaly**

**Prevalance:** 1:50-100000 live births

**Clinical Features:**
- Depends on the severity of the pathology and the magnitude of L-R shunt
- In milder forms only murmur and arrhythmia
- In severe forms cyanosis related to the extent of R-L shunting
- WPW Syndrome

[www.escardio.org/EAE](http://www.escardio.org/EAE)
Ebstein Anomaly-Echocardiography

- Increase in RV volume
- Paradoxical septal motion
- Increase in TV excursion
- Delay in TV closure compared to MV closure
- Decreased TV EF slope
Surgical Management

• Indications for intervention
  - Deteriorating FC (NYHA≥Class III)
  - Progressive cyanosis
  - Right heart failure
  - Paradoxic embolism
  - Recurrent supraventricular arrhythmia
  - Asymptomatic progressive cardiomegaly
# Indications for Intervention in Ebstein’s Anomaly

### Indications for Surgery

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class &gt;II or arrhythmias) or deteriorating exercise capacity measured by LVEF</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray</td>
<td>IIa</td>
<td>C</td>
</tr>
</tbody>
</table>

### Indications for Catheter Intervention

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>If cyanosis (oxygen saturation at rest &lt;90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (see text)</td>
<td>IIb</td>
<td>C</td>
</tr>
</tbody>
</table>
Surgery

- Tricuspid valve repair
- Tricuspid valve replacement
- Plication of the atrialized RV
- For high risk patients a bidirectional cavo-pulmonary connection (to reduce RV preload)
- Ablation of the accessory pathways
- Closure of PFO/ASD if present
Late Complications

- Reoperation of TV if TR persist
- Valve replacement may be necessary if bioprostheses fails or mechanic valve thrombosis
- Late arrhythmias may occur
- Complete heart block may occur
Prognosis and Management

Survival at 1 year 67% and 10 year 59%.

Survival after Tricuspid bioprosthesis: 10 yr 93% and 15 yr 81%. 94% in NYHA I/II.

Prognosis is worse if:
- NYHA FC III-IV,
- CTR > 65%
- atrial fibrillation,
- severe cyanosis,
- severe TR,
- functional rv area < %35
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Transposition of the Great Arteries (TGA)

- D loop
- L Loop
- RV follow the loop
  D loop RV on the right
  L loop RV on the left
L- TGA - Congenitally corrected TGA

Atrio-ventricular discordance
Ventriculo-arterial discordance

Associated Anomalies: (98%)
-VSD (75%)
-P or subP stenosis (75%)
-L sided (TV- Ebstein like)
valvular anomalies (75%)
-Complete AV block (2%/y)
**D- TGA- Complete TGA**

- Incidence 20-30/1000,000 live births.
- Without treatment 30% die within 1st week, 50% within 1st month, 70% in 6 months and 90% in first year.
- With current medical and surgical interventions 90% early and midterm survival.
- In complete Transposition the connection between atria and ventricles are normal.
- The connections between the ventricles and the great arteries are discordant.
D-TGA, Complete TGA
Surgery

- Most commonly done procedures were atrial switch operations:
- Blood redirected at atrial level with a baffle made of dacron or pericardium (Mustard)
- With atrial flaps (Senning) to achieve physiological correction
Atrial Switch Surgery
(Mustard, Senning procedures)
Consequences of Mustard

- Most patients reaching adulthood have NYHA I/II symptoms over the next 25 years.
- 50% develop moderate **systolic dysfunction** of the RV but only few present with CHF.
- 1/3rd have **severe systemic TR**.
- **Atrial flutter** arises in 20% by age 20.
- 50% patients have **sinus node dysfunction** by age 20.
- **Baffle leak or obstruction** can also occur.
Baffle leak
### Indications for Catheter Interventions in TGA patients after Atrial Switch

<table>
<thead>
<tr>
<th>Indication</th>
<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stenting should be performed in asymptomatic patients with baffle stenosis</td>
<td>I C</td>
</tr>
<tr>
<td>Stenting (covered) or device closure should be performed in symptomatic patients with baffle leaks and substantial cyanosis at rest or during exercise</td>
<td>I C</td>
</tr>
<tr>
<td>Stenting (covered) or device closure should be performed in patients with baffle leaks and symptoms due to L-R shunt</td>
<td>I C</td>
</tr>
<tr>
<td>Stenting (covered) or device closure should be considered in asymptomatic patients with baffle leaks with substantial ventricular volume overload due to L-R shunt</td>
<td>IIa C</td>
</tr>
<tr>
<td>Stenting should be considered in asymptomatic patients with baffle stenosis who require a PM</td>
<td>IIa C</td>
</tr>
<tr>
<td>Stenting may be considered in other asymptomatic patients with baffle stenosis</td>
<td>IIb C</td>
</tr>
</tbody>
</table>

ESC GUCH Guidelines 2010
## Indications for Surgical Interventions in TGA patients after Atrial Switch

### ESC GUCH Guidelines 2010

<table>
<thead>
<tr>
<th>Indication</th>
<th>Evidence Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valve repair or replacement should be performed in patients with severe</td>
<td>I</td>
</tr>
<tr>
<td>symptomatic systemic (tricuspid) AV valve regurgitation without significant</td>
<td></td>
</tr>
<tr>
<td>ventricular dysfunction (RVEF &gt;45%)</td>
<td></td>
</tr>
<tr>
<td>Significant systemic ventricular dysfunction, with or without TR, should</td>
<td>I</td>
</tr>
<tr>
<td>be treated conservatively or eventually with cardiac transplantation</td>
<td></td>
</tr>
<tr>
<td>LVOTO if symptomatic or if LV function deteriorates should be treated</td>
<td>I</td>
</tr>
<tr>
<td>surgically</td>
<td></td>
</tr>
<tr>
<td>In symptomatic pulmonary venous obstruction surgical repair (catheter</td>
<td>I</td>
</tr>
<tr>
<td>intervention rarely possible) should be performed</td>
<td></td>
</tr>
<tr>
<td>Symptomatic patients with baffle stenosis not amenable for catheter</td>
<td>I</td>
</tr>
<tr>
<td>intervention should be treated surgically</td>
<td></td>
</tr>
<tr>
<td>Symptomatic patients with baffle leaks not amenable for stenting should</td>
<td>I</td>
</tr>
<tr>
<td>be treated surgically</td>
<td></td>
</tr>
<tr>
<td>Valve repair or replacement should be considered for severe</td>
<td>IIa</td>
</tr>
<tr>
<td>asymptomatic systemic (tricuspid) AV valve regurgitation without significant</td>
<td></td>
</tr>
<tr>
<td>ventricular dysfunction (RVEF &gt;45%)</td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery banding in adult patients, to create septal shift, or as</td>
<td>III</td>
</tr>
<tr>
<td>left ventricular training with subsequent arterial switch, is currently</td>
<td></td>
</tr>
<tr>
<td>experimental and should be avoided</td>
<td></td>
</tr>
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</table>
Rastelli Procedure

• Procedure for TGA with VSD and pulmonary/subpulmonary stenosis
• Blood is redirected at ventricular level
• LV tunneled to Ao via VSD
• RV- PA via a valved conduit
Complications of the Rastelli Procedure

- RV- PA conduit stenosis causing exercise intolerance or RV angina
- Subaortic obstruction (LV-Ao tunnel) causing dyspnea or syncope
- Residuel VSD
Surgery

- Current practice is the **arterial switch** operation developed in 1980’s.
- Blood is redirected at the level of the great artery
- the morphological left ventricle becomes the subaortic ventricle the morphological
- right ventricle becomes the subpulmonic ventricle
Arterial Switch Operation
Consequences of Arterial Switch

- supra neopulmonary artery stenosis
- ostial coronary artery disease
- progressive neoaortic valve regurgitation
## Indication for Interventions in TGA patients after Arterial Switch

<table>
<thead>
<tr>
<th>Indications</th>
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<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stenting or surgery (depending on substrate) should be performed for coronary artery stenosis causing ischaemia</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Surgical repair of RVOTO should be performed in symptomatic patients with RV systolic pressure &gt; 60 mmHg (TR velocity &gt; 3.5 m/s)</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Surgical repair of RVOTO should be performed regardless of symptoms when RV dysfunction develops (RVP may then be lower)</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Surgical repair should be considered in asymptomatic patients with RVOTO and systolic RVP &gt; 80 mmHg (TR velocity &gt; 4.3 m/s)</td>
<td>IIA</td>
<td>C</td>
</tr>
<tr>
<td>Aortic root surgery should be considered when the (neo-)aortic root is &gt; 55 mm, providing average adult stature (for aortic valve replacement for severe AR see guidelines for AR)</td>
<td>IIA</td>
<td>C</td>
</tr>
<tr>
<td>Stenting or surgery (depending on substrate) should be considered for peripheral PS, regardless of symptoms. If &gt; 50% diameter narrowing and RV systolic pressure &gt; 50 mmHg and/or lung perfusion abnormalities are present</td>
<td>IIA</td>
<td>C</td>
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Single Ventricle Physiology and Fontan Circulation

- Tricuspid atresia
- Hypoplastic right heart syndrome variants, e.g. pulmonary atresia with intact ventricular septum variants
- Hypoplastic left heart syndrome variants, including mitral atresia
- Double-inlet LV
- Double-inlet RV
- Extreme forms of unbalanced complete AV septal defects
- Single ventricle with undefined morphology.

Biventricular repair can not be done
Tricuspid Atresia Single Ventricle
## Indication for Intervention in Single Ventricles

<table>
<thead>
<tr>
<th>Special considerations and indications</th>
<th>Class^a</th>
<th>Level^b</th>
</tr>
</thead>
<tbody>
<tr>
<td>Only well-selected patients after careful evaluation [low pulmonary vascular resistances, adequate function of the AV valve(s), preserved ventricular function] should be considered candidates for a Fontan operation</td>
<td>IIA</td>
<td>C</td>
</tr>
<tr>
<td>Patients with increased pulmonary blood flow—unlikely at adult age—should be considered for PA banding or tightening of a previously placed band</td>
<td>IIA</td>
<td>C</td>
</tr>
<tr>
<td>Patients with severe cyanosis, with decreased pulmonary blood flow without elevated PVR, should be considered for a bidirectional Glenn shunt</td>
<td>IIA</td>
<td>C</td>
</tr>
<tr>
<td>Heart transplantation and heart–lung transplantation should be considered when there is no conventional surgical option in patients with poor clinical status</td>
<td>IIA</td>
<td>C</td>
</tr>
</tbody>
</table>

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Glenn Procedure

- Palliative procedure for cyanotic patients or as a step to Fontan procedure or corrective surgery (e.g. single ventricle)
- Goal: to improve pulmonary blood flow
- Establishes a direct connection between SVC and right PA (directs half of the blood volume directly to the lung without the assistance of the ventricle)
- Does not create volume overload
- The venous return is under low pressure low risk for pulmonary vascular obstructive disease
- Performed at 3-8 months of age
Glenn Shunt

Bi-directional Glenn Shunt
Temporary surgical procedure as an intermediate procedure for hearts with only one usable ventricle (in this illustration Tricuspid Atresia)
Fontan Circulation

- Used when biventricular connections are not possible, the two functioning ventricles cannot be effectively established (e.g. double-inlet single ventricle, tricuspid atresia)
- Directs the blood directly to the lungs without assistance of the ventricle
Fontan  Total Cavo-Pulmoner Anastomosis
Fontan Prognosis

• 10 year survival is 60-70%
• Late term complications include
  - Atrial flutter or fibrillation
  - Right atrial thrombus
  - Obstruction of the Fontan circuit
  - Ventricular dysfunction
  - Protein loosing enteropathy
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Contributions Omaç Tüfekçioğlu M.D.