D-transposition of the great arteries

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No disclosures
Complete transposition of the great arteries is a congenital cardiac anomaly in which the aorta arises from the morphological right ventricle, and the pulmonary artery arises from the morphological left ventricle (ventriculoarterial discordant connection).
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Prevalence

- Prevalence per 10,000 live births: 2.3
- Ratio male / female: 2.9*

*erratum „LL 2:3“

Prevalence of congenital heart defects in newborns in Germany: Results of the first registration year of the PAN Study (July 2006 to June 2007)

Lindinger, A., G. Schwedler, and H.W. Hense, Klin Padiatr; 222: 321-326
Genetics and risk factors

There might be an increased prevalence in infants with
• diabetic mothers
• mothers with alcohol abuse during pregnancy
• mothers with malnutrition.

But there is
• no specific genetic pattern
• no association with the more common genetic disorders

Exception: Xlinked Heterotaxy (Mutations in Zic3)

Usually the aortic valve is positioned anteriorly and to the right {S,D,D} with a pulmonary-mitral continuity.

The clinical anatomy of transposition
Robert H. Anderson, Paul M. Weinberg
Cardiol Young 2005; 15 (Suppl. 1): 76–87
The pulmonary and systemic circulations function in parallel, so

- Oxygenated pulmonary venous blood returns via left atrium and ventricle to the pulmonary vascular bed
- Deoxygenated systemic venous blood returns via right atrium and ventricle to the systemic circulation.

This leads to

- deficient oxygen supply to the tissues
- excessive left ventricular volume workload.

Survival is not possible unless mixing of oxygenated and deoxygenated blood occurs.
Simple type 75%
(intact ventricular septum, or a ventricular septal defect is so small with no haemodynamic significance*)

Complex type 25%
- large VSD 20%
- Obstructive outflow 5%

*The clinical anatomy of transposition
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LVOTO - subpulmonary stenosis

The clinical anatomy of transposition
Robert H. Anderson, Paul M. Weinberg
Cardiol Young 2005; 15 (Suppl. 1): 76–87
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Coronary anatomy

Moss and Adams’ Heart Disease in Infants, Children and Adolescents, 6th ed. Philadelphia: Lippincott, Williams & Wilkins; 2001
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Coronary anatomy

Two Coronary Ostia

Single Coronary Ostium

Intramural Coronary Artery

Coronary Artery Pattern and Outcome of Arterial Switch Operation for Transposition of the Great Arteries

Circulation. 2002;106:2575-2580
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Physical findings

The clinical course of an infant with TGA is cyanosis - not reversible with oxygen supply – and heart failure with early death!

Second heart sound is loud and single
Heart murmur depends on VSD, LVOTO
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Diagnostic goals

- Confirm diagnosis
- Description of anatomy (incl. Coronary anatomy)
- Concomitant heart defects (VSD, LVOTO, RVOTO, CoA)
- Rating of mixing via PFO, PDA (VSD)
- Assess and quantify cyanosis
- Judgement of effectiveness of prostaglandin E1 therapy
- For indication of atrial septostomy

Echocardiography
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Echocardiography

Subcostal sagittal view

Parasternal short-axis

William T. Mahle, Javier H. Gonzalez, Joseph Kreeger, Gerald Marx, Gul Duldani, Norman H. Silverman

Echocardiography of transposition of the great arteries
Cardiology in the Young (2012), 22, 664–670
Parasternal long-axis demonstrates the origin of the right coronary artery

Parasternal short-axis image demonstrating the origin of the left coronary artery

William T. Mahle, Javier H. Gonzalez, Joseph Kreeger, Gerald Marx, Gul Duldani, Norman H. Silverman

Echocardiography of transposition of the great arteries
Cardiology in the Young (2012), 22, 664–670
Parasternal short-axis image demonstrating the intramural course of the left coronary artery

Apical image demonstrating the circumflex coronary artery passing posterior to the pulmonary annulus

William T. Mahle, Javier H. Gonzalez, Joseph Kreeger, Gerald Marx, Gul Duldani, Norman H. Silverman

**Echocardiography of transposition of the great arteries**
Cardiology in the Young (2012), 22, 664–670
Parasternal short-axis view demonstrating bowing of the ventricular septum towards the left ventricle (LV) suggesting elevated right ventricular (RV) pressure relative to LV.

M-mode measurements can help to estimate the left ventricular mass

William T. Mahle, Javier H. Gonzalez, Joseph Kreeger, Gerald Marx, Gul Duldani, Norman H. Silverman

Echocardiography of transposition of the great arteries
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Diagnostic methods

- Pulse oxymetry: level of cyanosis
- Blood gas analysis: additional information about circulatory failure
- Chest X-ray: evaluation of lung condition
- ECG: reference for postoperative examination
Cardiac catheterisation

Usually not necessary, but indicated
- in complex cases
- in undefined coronary anatomy
- in case of late presentation and unclear hemodynamics
Usually not indicated, but …

In case of late presentation (with / without LV retraining) MRI can provide helpful information about left ventricular mass and function.
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First line therapy: improve mixing

Prostaglandin E infusion

- start with 10 – 20 (50) ng/kg/min, if possible
- decrease to 5-10 ng/kg/min

- check
  - blood gas analysis
  - echocardiography (PDA, atrial mixing, abdominal perfusion)
Inadequate intracardiac mixing due to a restrictive foramen ovale will not improve solely on PGE.

The markedly increased pulmonary blood flow from the PDA may lead to deleterious left atrial hypertension, pulmonary congestion, and low cardiac output.
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atrio septostomy

indication

- in case of restricted atrial septal flow
- no sufficient improvement despite prostaglandin E infusion
- to avoid, reduce or terminate prostaglandin E infusion (e.g. NEC, renal insufficiency, delayed corrective surgery)
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surgery

1. choice
arterial Switch operation

2. choice
atrial Switch operation

D-TGA/ IVS

D-TGA&VSD

D-TGA&VSD&LVOTO

Rastelli REV
Nikaidoh-Bex
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Arterial switch operation

30d-mortality rate (EACTS)
Simple TGA  3-4%
TGA & VSD  7%

Orphanet Journal of Rare Diseases 2008,3:27
### Guideline d-TGA

**Arterial switch operation**

<table>
<thead>
<tr>
<th>Long-Term Post-Operative Sequelae</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supravalvular pulmonary stenosis*</td>
<td>~10%</td>
</tr>
<tr>
<td>Supravalvular aortic stenosis*</td>
<td>~5%</td>
</tr>
<tr>
<td>Neoaortic root dilation</td>
<td>Nearly universal</td>
</tr>
<tr>
<td>Neoaortic regurgitation</td>
<td>Most (moderate or severe in &lt;10%)</td>
</tr>
<tr>
<td>Asymptomatic coronary occlusion</td>
<td>2%-7%</td>
</tr>
<tr>
<td>Sudden cardiac death</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>2%-10%</td>
</tr>
<tr>
<td>Aortic dissection or rupture</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

Aortic and pulmonary artery vessel wall and sinus samples were taken from 20 untreated human heart specimens with transposition of the great arteries and 9 age-matched, normal, postmortem human heart specimens.

In the pulmonary artery and sinus of untreated transposition of the great arteries, there is a *dedifferentiation of smooth muscle cells* with increasing age that we could not correlate to altered flow.

Pulmonary artery remodeling in transposition of the great arteries: Relevance for *neoaortic root dilatation* Shirin Lalezari, Mark G. Hazekamp, Margot M. Bartelings, Paul H. Schoof, Adriana C. Gittenberger-de Groot J Thorac Cardiovasc Surg 2003;126:1053-1060
The Mustard procedure and Senning procedure are identical except that the baffle is constructed from atrial tissue in the Senning and from pericardium in the Mustard procedure.

Orphanet Journal of Rare Diseases 2008,3:27

Late problems after atrial switch operation

- Baffle leaks
- Baffle obstruction
- Tricuspid valve insufficiency
- RV-dysfunction
- Residual lesions (VSD, LVOTO)
- Arrhythmia

Late outcome of Senning and Mustard procedures for correction of transposition of the great arteries

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TGA, VSD, LVOTO

- Rastelli
- REV
- Nikaidoh-Bex

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Rastelli

REV
Reparation a l´étage ventriculaire

Orphanet Journal of Rare Diseases 2008,3:27
two main problems: subaortic stenosis
RV—PA conduit or connection stenosis

J Thorac Cardiovasc Surg 2008;135:331-338
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Bex-Nikaidoh in TGA, VSD, LVOTO

Orphanet Journal of Rare Diseases 2008, 3:27
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Early and late problems after aortic root translocation

- Sudden cardiac death
- Coronary distortion/kinking
- Aortic valve insufficiency
- Pulmonary insufficiency

In case of pulmonary re-rooting
- Arrhythmia, AV-Block
- Mitral valve insufficiency