

CLINICAL PRACTICE GUIDELINE

Guideline coverage includes NICU KEMH, NICU PCH and NETS WA

Transposition of the Great Arteries (TGA)

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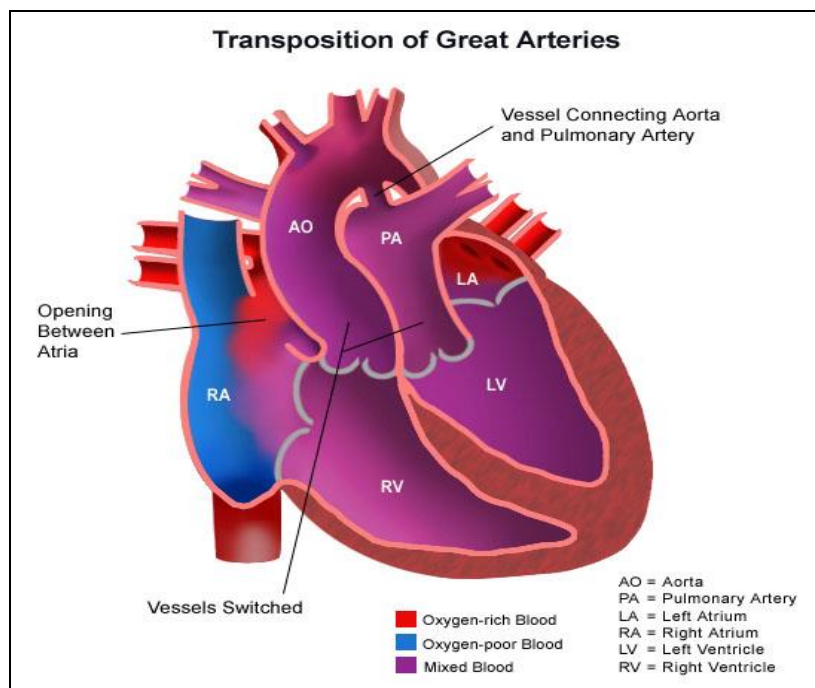
Transposition of the great arteries (TGA) is a common congenital cardiac anomaly and accounts for 4-5% of all congenital heart disease. Incidence of 0.2-0.4/1000 live births and it is more prevalent in boys than girls, ratio of 3:1. If left untreated most babies will die during the first year of life.

Anatomy

In a simple dextro-TGA the aorta and main pulmonary artery [PA] are switched such that the aorta emerges from the right ventricle (RV) anteriorly and the main PA from the left ventricle (LV) posteriorly.

There are other types of TGA including levo-TGA where the ventricles are also transposed, known as the congenitally corrected TGA. TGA may be associated with other cardiac anomalies such as VSD, ASD, DORV, pulmonary stenosis with VSD and left ventricular outflow tract obstruction (LVOTO).

Dextro-TGA with/without VSD and management will be discussed here.



Physiology of the Lesion

The pulmonary and systemic circulations are in parallel rather than in series. This may lead to life-threatening hypoxaemia in the systemic circulation and survival depends upon the presence of one or more mixing points (ASD, VSD, PDA) between the two circulations. The amount of mixing greatly influences the SaO₂ and severity of the clinical picture.

Clinical Presentation

Newborns with TGA/intact ventricular septum (IVS) with a small patent foramen ovale (PFO) or PDA have severe cyanosis on day 1 of life; sometimes with cardiovascular collapse as the PDA shuts. TGA should be suspected in any newborn with cyanosis not responsive to O₂ therapy.

Those with TGA/VSD or TGA/IVS with a large ASD or PDA have better mixing and hence higher pO₂, but they also have a greater tendency to develop congestive cardiac failure, presenting as tachypnoea and respiratory distress.

Genetic/Syndromic Associations

There is no genetic predisposition TGA and it is typically not associated with other non-cardiac anomalies.

Management of a Neonate with TGA

Initial Management at Delivery when Antenatally Diagnosed

- A Neonatal Management Plan (MR409.90) should be in place.
- Usual postnatal resuscitative measures.
- Commence PGE1 ([Alprostadil](#)) and contact cardiologist.
- Make arrangements for transfer to PCH when stable.

Presentation with Cyanosis/Acidosis/Cardiovascular Collapse

- Airway + Breathing:
 - Most likely will require mechanical ventilation (especially if transporting on PGE1 infusion).
 - Aim for SaO₂ > 70% and normocarbia (note: the baby may have a low PaCO₂ to compensate for metabolic acidosis).
- Circulation:
 - Gain PIV access - peripheral (avoid umbilical vessels unless required in emergency as most likely will be required for atrial septostomy).
 - Peripheral Arterial line helpful but not essential - peripheral (see above).
 - Commence PGE1 as soon as possible at 10-50ng/kg/min.
 - If acidotic and shocked, give normal saline fluid boluses in aliquots of 10mL/kg up to a total of 20mL/kg total.
 - Consider inotropes after 20mL/kg of saline if ongoing hypotension or acidosis. Start with dopamine 10 mcg/kg/min.
- Give IV Penicillin and Gentamicin if sepsis possible.
- Elective/semi elective [intubation](#) is best performed with Morphine or Fentanyl, Atropine and Suxamethonium as per usual Neonatal Directorate guideline. Ongoing opioid sedation will be required but not usually paralysis.
- Maintain normothermia and normoglycaemia
- Communicate with cardiology/NETS WA early.

Investigations

- Blood gas - normal, compensated or uncompensated metabolic acidosis.
- Routine Bloods: FBC, Coagulation profile, Electrolytes and creatinine, Blood glucose, Liver Function Tests, Ca/Mg, CRP, Blood culture.
- Blood 'Group and hold'

- Chromosomal analysis - not usually required.
- CXR - The typical 'egg-on-side' may not be obvious early. Check placement of tubes/lines.
- ECHO details presence of VSD, PFO or ASD and direction of shunting, PDA status and shunting, anatomy of the coronary arteries, aortic arch and it's orientation, LVOT, presence of pulmonary stenosis, RV size.
- Head USS - As a pre op baseline especially in those who present shocked.
- Renal USS - Not usually required.

Preoperative Management

CVS

- Continue PGE1: dose varies according to PDA status and results of balloon atrial septostomy.
- Blood gases should be followed closely and should quickly normalise.
- Maintain mean BP mean in the upper normal range.
- Inotropic support maybe required e.g. Dopamine 10-20 mcg/kg/min. Further inotrope use depends on ECHO findings and clinical picture.

Respiratory

- Ventilation or nasal CPAP depends on the ongoing clinical picture.
- SIPPV+VG (4-6ml/kg) with 5 cmH₂O PEEP. A higher PEEP may be required if there are signs of pulmonary oedema/over-circulation.
- Aim for normocarbia.
- FiO₂ to keep the SaO₂ > 70%. Over oxygenation may cause increased pulmonary shunting.
- Consider extubation once blood gases are normalised, PGE1 is very low or not needed (adequate atrial septostomy/mixing).

Fluids/Nutrition

- Normal maintenance fluid for age. Fluid restrict in heart failure or fluid overloaded (Frusemide may be required).
- Monitor electrolytes and correct accordingly.
- Record and monitor urine output - should be > 1 mL/kg/hr.
- Commence feeds when cardio-metabolic stability is achieved.

Sepsis

- Antibiotics are continued until cultures are clear and CRP has normalised.

Haematology

- Correct coagulopathy/low platelet count.
- Hb is usually maintained above 120g/L

Neuro

- Usually normal unless previously shocked.

Lines

- Avoid umbilical vessels as detailed above.
- An arterial line is helpful prior to septostomy and/or unstable patients.

Balloon Atrial Septostomy (Rashkind Procedure)

Those patients with an intact ventricle and a restrictive PFO and poor mixing will require an urgent balloon atrial septostomy under echocardiographic guidance.

The procedure involves catheterisation of the umbilical or femoral vein; the catheter is guided into the right atrium and through the PFO into the left atrium. The catheter balloon is inflated and pulled back through the PFO to enlarge it (usually > 4mm).

Preparation for Balloon Atrial Septostomy

- Check coagulation profile/platelet count and correct any abnormalities.
- Ensure 'Group and Hold' has been sent.
- All patients will need some degree of sedation and often ventilation.
- The cardiologist should gain consent from the parents.

Following an adequate atrial septostomy PGE1 can be stopped. Occasionally the PGE1 is required for a short period after the septostomy.

Possible Pre-Operative Complications

- Multi organ dysfunction can occur in shocked patients causing renal, hepatic and cardiac impairment, coagulopathy, seizures and risk of NEC.

Usual Operative Management/Treatment Options

The treatment of choice for simple TGA is the arterial switch operation (ASO) usually in the first 2 weeks (5-10 days is optimal) of life once the pulmonary vascular resistance has dropped (to avoid pulmonary hypertensive crisis).

Post-Op outcomes

- Post op recovery is usually uneventful.
- The hospital mortality for ASO performed is approximately 0.9% for TGA/IVS and 4.1% for TGA/VSD.
- Late death after ASO has been rare with survival at 10 and 15 years as 93% and 86% respectively.
- Long term cardiac morbidity due to arrhythmias, valve dysfunction, and myocardial ischaemia may occur. Rhythm disturbance incidence: AV node dysfunction (4.4%), heart block requiring pacemaker (1.7%) (all had VSD) and SVT (5%) at late follow-up. Long term stenosis or occlusion of the coronary arteries may occur (3-7%).
- Most newborns undergoing ASO for TGA have a low risk of long-term neurodevelopmental impairment. Routine post op MRI of the brain is performed and all neonates with ASO and are enrolled in long term neurodevelopmental follow up.
- Antibiotic prophylaxis is required for life.

References

1. Nichols, D.G.; Ungerleider, R.M.; Spevak, P.J.; Greeley, W.J.; Cameron, D.E.; Lappe, D.G.; Wetzel, R.C., *Critical heart disease in infants and children*. Second ed.; Mosby: USA, 2006.
2. Horrox, F., *Manual of neonatal and paediatric heart disease*. First ed.; Whurr Publishers: Gateshead, Tyne and Wear, UK, 2002.
3. Koenig, P.; Hijazi, Z.M.; Zimmerman, F., *Essential pediatric cardiology*. First ed.; McGraw-Hill Companies, Inc: USA, 2004.

Related WNHS policies, procedures and guidelines

Neonatal Clinical Guideline - [Intubation](#)

Neonatal Medication Protocol - [Alprostadil](#)

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