GUIDELINE

Transposition of the Great Arteries (TGA)

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

Aim

Outlines the presentation and management of Transposition of the Great Arteries (TGA) in the neonatal population.

Risk

Failure to follow this guideline may lead to adverse outcomes for the infant.

Background

Transposition of the great arteries 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

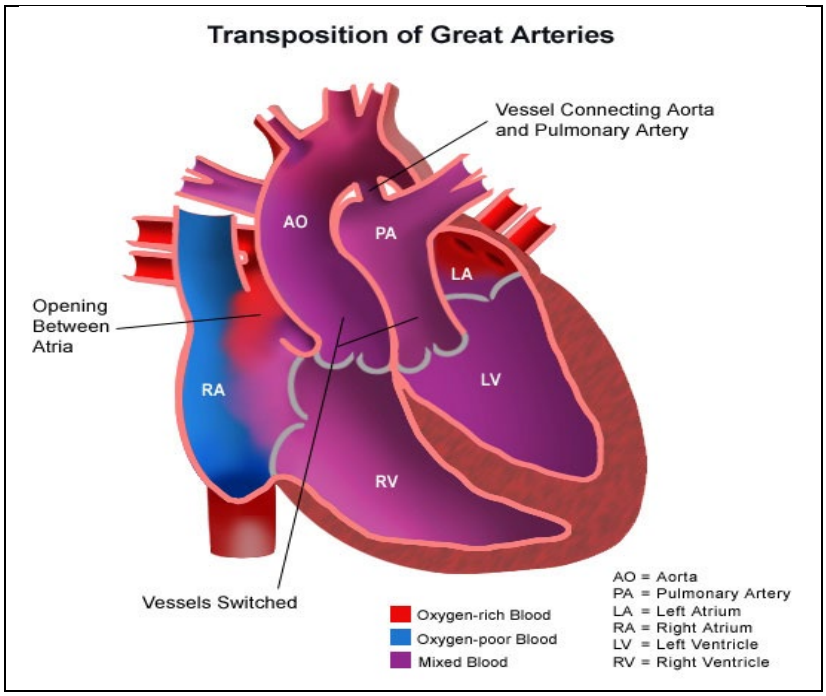
In a simple dextro-TGA (d-TGA) the aorta and main pulmonary artery [PA] are switched such that the aorta emerges from the right ventricle (RV) anteriorly and the main PA from the left ventricle (LV) posteriorly.

The other type of TGA is levo-TGA (l-TGA) where the ventricles are also transposed, known as the congenitally corrected TGA (cc-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 SaO_2 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 considered in any newborn with cyanosis not responsive to O_2 therapy.

Those with TGA/VSD or TGA/IVS with a large ASD or PDA have better mixing and hence higher paO_2, 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 to 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.

- Contact cardiologist.
- Commence PGE1 (Alprostadil) as directed by the Neonatal Management Plan or the cardiologist, or if in doubt.
- Make arrangements for transfer to PCH when stable.

Presentation with Cyanosis/Acidosis/Cardiovascular Collapse

- Airway + Breathing:
  - Consider mechanical ventilation (especially if transporting on PGE1 infusion).
  - Aim for SaO_2 > 70% (ideally 75-85%)
  - In setting of cyanosis due to low pulmonary blood flow, PaCO2 in the low-normal range promotes pulmonary blood flow (note: the baby may have a low $PaCO_2$ to compensate for metabolic acidosis).
- Circulation:
  - Gain intravenous access x2 - peripheral
    - avoid umbilical vessels unless required in emergency as umbilical access will most likely will be required for balloon atrial septostomy (BAS)
  - Peripheral arterial line access is helpful but not essential
  - Commence PGE1 as soon as possible at 10-50ng/kg/min (dose guided by cardiologist).
  - If acidotic and shocked, consider 0.9% Sodium Chloride fluid boluses in small aliquots of 5-10mL/kg up to a total of 20mL/kg.
  - Consider inotropes after 20mL/kg of 0.9% Sodium Chloride if ongoing hypotension or acidosis or earlier if shock is cardiogenic in origin.
- Give IV Benzylpenicillin and Gentamicin if sepsis possible.
- Elective/semi elective intubation is best performed with Fentanyl, Atropine and Suxamethonium as per usual Neonatal Intubation Guideline. See Medication Monographs for prescribing and dosage.
- 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’.

  - Request crossmatch x 1 unit urgently if suspected to need Cardiac Management of Balloon Atrial Septostomy
  - 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 degree of mixing through PDA and timing/result of BAS.
- Blood gases should be followed closely and should quickly normalise following a successful BAS. The development of increasing metabolic acidosis/ raised lactate along with increased SaO_2 (>90%) may reflect pulmonary over circulation.
- Maintain mean BP in the upper normal range to encourage pulmonary blood flow if the patient's saturations remain <70%.
- Inotropic support may be required, and use depends upon Echo findings and clinical picture.

Respiratory

- Ventilation or nasal CPAP depends upon the ongoing clinical picture.
- SIPPV+VG (4.5-6ml/kg) with 5-6cmH_2O PEEP. A higher PEEP may be required if there are signs of pulmonary oedema/over-circulation.
- Aim for normocarbia.
- FiO_2 to keep the SaO_2 75-85%. Over oxygenation may cause pulmonary over-circulation. Saturation targets may be lowered at the discretion of the Cardiologist.
- Persistent desaturation despite septostomy/ PDA may indicate PPHN and may need treatment accordingly.
- Consider extubation once blood gases are normalised and PGE1 dose 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 and slowly grade up when cardio-metabolic stability is achieved.

Sepsis

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

Haematology

- Correct coagulopathy/low platelet count.
- Haemoglobin should be maintained above 120g/L (level for cyanotic heart disease)

Neurology

- Usually normal unless previously shocked.

Intravenous and Arterial Lines

- Avoid umbilical vessels as detailed above until septostomy performed or a document decision has been made that a septostomy is not required.
- An arterial line is helpful prior to septostomy and/or unstable patients.

Balloon Atrial Septostomy (Rashkind Procedure)

Those patients with an intact ventricular septum and a restrictive PFO causing poor mixing will require an urgent BAS 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).

For further details on the procedure see Cardiac Management of Balloon Atrial Septostomy. Following an adequate atrial septostomy PGE1 can often be stopped. Occasionally the PGE1 is required for a short period after the septostomy.

Possible Pre-Operative Complications

- Multiorgan dysfunction can occur in shocked patients causing renal, hepatic and cardiac impairment, coagulopathy, seizures and risk of NEC.
- Persistent pulmonary hypertension of the newborn (PPHN) sometimes occurs and requires treatment with nitric oxide (NO).

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-Operative Outcomes

- Post-operative 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 should be discussed with cardiology.

Related CAHS internal policies, procedures and guidelines

Neonatology Guideline

- Intubation
- Cardiac Management of Balloon Atrial Septostomy
- Persistent pulmonary hypertension of the newborn (PPHN)

Neonatal Medication Protocol

King Edward Memorial Hospital - Neonatal Medication Protocols (health.wa.gov.au)

References and related external legislation, policies, and guidelines

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.

This document can be made available in alternative formats on request.

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