



SECTION: 14 NEONATAL CARDIAC CONDITIONS

TRANSPOSITION OF THE GREAT ARTERIES (TGA)

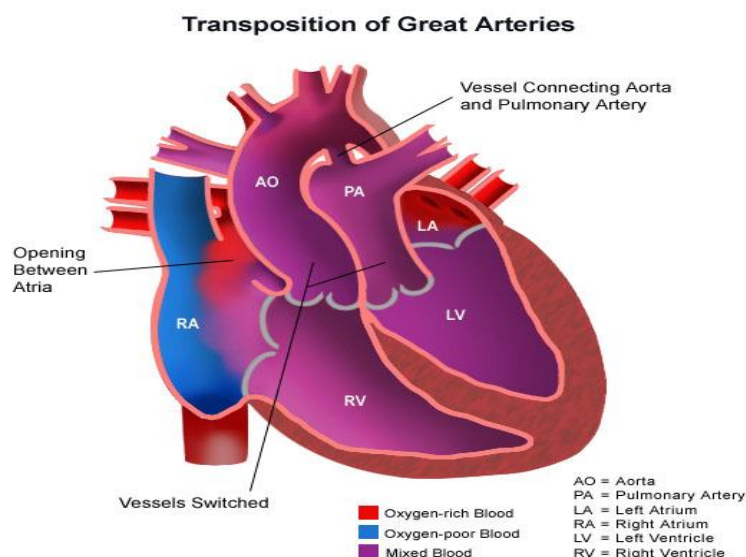
Transposition of the great arteries (TGA) is the second most common congenital cardiac anomaly encountered in early infancy and accounts for 4-5% of all congenital heart disease. It has a reported incidence of 0.2-0.4/1000 live births. It is found to be more prevalent in boys than girls in a ratio of 3:1. If left untreated, approximately 90% will die in the first year of life.

ANATOMY

In TGA the great vessels that arise from the heart (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. This can further be described as atrioventricular (AV) concordance with ventriculoarterial (VA) discordance. If no other cardiac defect is present, this is termed simple transposition, or d-TGA (where 'd' [dextrotransposed] denotes the location of the aorta). If there are other defects present, then there is a complex transposition.

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

The more common d-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 arterial in the systemic circulation and survival depends upon the presence of one or more crossover or mixing points (ASD, VSD, PDA) between the two circulations. The amount of mixing of caval and pulmonary venous blood 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 acidosis and 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 within 2 weeks of birth.

GENETIC/ SYNDROMIC ASSOCIATIONS

There is no apparent genetic predisposition to this congenital heart disease and it is typically not associated with other anomalies.

MANAGEMENT OF A NEONATE WITH TRANSPOSITION OF THE GREAT ARTERIES (TGA)

INITIAL MANAGEMENT AT DELIVERY WHEN ANTENATALLY DIAGNOSED:

- Most simple TGAs diagnosed antenatally will be transferred in utero to Melbourne.
- Usual postnatal resuscitative measures.
- Admit to SCN3, assess, commence PGE1/ Alprostadil and contact cardiologist early.
- Make arrangements for transfer to ward 6B when stable and safe to do so.

PRESENTATION WITH CYANOSIS/ACIDOSIS/CV COLLAPSE

1. Airway + Breathing:

- Most likely will require mechanical ventilation (especially if transporting on PGE1 infusion).
- Use pressures/ FiO₂ to keep normocarbida (though patent may breath up and ↓pCO₂ to compensate for metabolic acidosis) and SaO₂ >70%.

2. Circulation:

- Gain IV access – peripheral (avoid umbilical vessels unless required in emergency as most likely required for atrial septostomy).
- Arterial line helpful but not essential – peripheral (avoid umbilical vessels unless required in emergency as most likely required for atrial septostomy).
- Commence PGE1/ Alprostadil as soon as possible at 50ng/kg/min.
- If acidotic and shocked, give normal saline fluid boluses in aliquots, maximum of 10mL/kg with each. Give up to 20mL/kg total and if requiring more than this, discuss with consultant as may be detrimental.
- Sometimes when patient is shocked and acidotic, heart requires support with inotropes. Start with dopamine 10mcg/kg/min. (If no central access, dobutamine up to 10mcg/kg/min may be used peripherally, but watch infusion site carefully).

3. Give IV amoxicillin and gentamicin if sepsis possible.

4. Use small amount of sedation for intubation and as ongoing infusion (50mcg/kg bolus flowed by 10mcg/kg/hr) so as not to compromise cardiac function further. Paralysis is not normally necessary or desirable.

5. Maintain normothermia/normoglycaemia

6. Consider other causes shock:

- Sepsis
- Other cardiac disease
- Metabolic disease
- Intra-abdominal pathology such as volvulus
- Causes of bleeding such as trauma/ NAI/ Vit K deficiency

Communicate with cardiology/ NETS WA early.

INVESTIGATIONS

- **Gas** – Maybe normal, but if presenting in shock, may show severe metabolic/ lactic acidosis which may be partially compensated with a low pCO₂.
- **Routine Bloods:**
 - **FBC** – should have normal Hb, plt may be ↓ if period of hypoperfusion/ shock.
 - **Coags** – may be deranged (particularly INR) if period hypoperfusion/ shock.
 - **U+E** – electrolytes should be normal, but creatinine may be elevated if period of hypoperfusion/ shock.
 - **LFTs** – transaminases may be elevated if period hypoperfusion/ shock.
 - **Ca/Mg** – any patient in shock or when 22q11 deletion possible (eg. IAA). Ionised Ca on gas most helpful.
 - **CRP** – check for evidence sepsis.
 - **Blood culture** – if sepsis possible.
 - **G+H**
- **Chromosomal analysis** – not usually required as not usually associated with syndrome.
- **CXR** – Initially is usually normal. If there is a VSD and the patient is presenting later with signs of heart failure, there may be cardiomegaly and pulmonary plethora. The typical 'egg-on-side' is only noted at a later date when the narrow mediastinum is teamed with biventricular dilatation. Check placement of tubes/ lines.
- **Echo** – Is the gold standard for diagnosis of TGA in the newborn period and usually provides excellent detail of the surgically relevant features. Particular features that are assessed are: presence of VSD, PFO or ASD and direction of shunting, PDA and shunting, anatomy of the coronary arteries, aortic arch and it's orientation, LVOT, presence of pulmonary stenosis, RV size. It can then be used to aid balloon atrial septostomy if required.
- **Head USS** – As a baseline, and is particularly important for those who present unwell/ shocked. Should preferably be performed prior to theatre.
- **Renal USS** – Not usually required.

PREOPERATIVE MANAGEMENT

CVS

- Continue PGE₁/ Alprostadil until assessment by cardiologist. The dose may be decreased or stopped following balloon atrial septostomy, if adequate mixing achieved, on the advice of the cardiologist.
- The gas should be followed closely, and should normalise:
 - hourly initially until patient stabilising and metabolic acidosis improving.
 - then 2 hourly until metabolic acidosis/ lactate normalised.
 - then 4 hourly unless clinical deterioration.
- BP mean should be kept >45mmHg (unless otherwise stated by consultant or cardiologist).
- When there has been acidosis/ cardiovascular collapse, inotropic support of the heart is sometimes required. First choice is dopamine 10-20mcg/kg/min. If a further inotrope is required, then adrenaline or milrinone may be considered.

RESPIRATORY

- Most patients having presented cyanotic/ acidotic/ shocked/ on prostaglandins will initially be mechanically ventilated.



- Normal SIPPV+VG with normal term neonatal settings for PEEP and pressures should be used. A higher PEEP may be required if there are signs of pulmonary oedema in the case of later presentation with VSD.
- Patient should be ventilated to normocarbica (though as mentioned before, may breath up to hypocarbica to compensate for metabolic acidosis until it has resolved).
- The minimum FiO₂ to keep the SaO₂ >70% should be used. Extra O₂ should not be used as this may cause pulmonary dilatation and increased pulmonary shunting.
- Often, once adequate atrial septostomy has been performed, and off PGE₁, the patient is able to be extubated.

FLUIDS/ NUTRITION

- The patient should be given normal amount of fluid for age. However, if the patient is presenting in cardiac failure, fluids may need to be restricted. 10% dextrose + 0.22% saline should be used with potassium added as required.
- Watch electrolytes and correct accordingly.
- If there is evidence of cardiac failure, frusemide may be required.
- Urine output should be recorded hourly initially, and should be >1mL/kg/hr.
- Initially the patient should be NBM and this sometimes needs to be for a few days if there has been a period of acidosis and shock (risk of NEC).
- Once adequate mixing has been established and the patient is stable, feeds can usually be introduced.

SEPSIS

- As sepsis is so common in neonates, and not to be missed, most will be given at least 48 hours IV amoxicillin and gentamicin until cultures are back.

HAEMATOLOGY

- Correct coagulopathy/ low platelet count as necessary.

NEURO

- Sedation with low dose morphine is usually all that is required.
- Muscle relaxation is rarely required or desired.

LINES

- Umbilical vessel catheterisation prior to atrial septostomy should be avoided unless other access cannot be gained. Usually the cardiologist is happy to leave a UVC in once septostomy is completed.
- In the acidotic/ shocked patient an arterial line is helpful and should be placed peripherally prior to septostomy. Once the patient is stable it should be removed.

BALLOON ATRIAL SEPTOSTOMY (RASHKIND PROCEDURE)

Those with TGA/IVS that are found to have a restrictive PFO and poor mixing on echocardiogram will require an urgent balloon atrial septostomy under echocardiographic guidance by the cardiologist on 6B.

The procedure involves catheterisation of the umbilical vein or femoral vein and feeding it up to the right atrium and through the PFO into the left ventricle. A balloon is then inflated and then pulled back through the PFO to enlarge it. Following the procedure, the cardiologist then determines that the defect created is adequate (usually at least 4mm) and that the margins are clear. The heart is also scanned for any iatrogenic damage.

PREPARATION FOR BALLOON ATRIAL SEPTOSTOMY

- Check coagulation profile/ platelet count and correct any abnormalities.
- Ensure G+H has been sent.
- The patient will be required to be quiet, but not heavily sedated/ paralysed. Most will be ventilated, so this can be easily achieved with a small bolus of morphine followed by and infusion.
- The cardiologist should gain consent from the parents.

If the atrial septostomy has provided adequate mixing, then the cardiologist will likely suggest stopping PGE1. Occasionally SaO₂ drop and the PGE1 requires recommencing.

POSSIBLE PRE-OPERATIVE COMPLICATIONS

- If there has been acidosis and cardiovascular collapse, there may be impairment of other organ function causing renal impairment, cardiac impairment, coagulopathy, seizures and risk of NEC.
- Once good mixing has been achieved with PGE1/ atrial septostomy, further pre-operative complications are rare.

USUAL OPERATIVE MANAGEMENT/ TREATMENT OPTIONS

The treatment of choice for simple TGA is the arterial switch operation (ASO) (*see protocol 'Other Cardiac Procedures' number 1*) usually in the first 2 weeks (5-10 days is optimal) of life once the pulmonary vascular resistance has dropped somewhat (to avoid pulmonary hypertensive crisis).

This is not presently offered at PMH, so transfer is necessary to another unit such as Royal Children's, Melbourne or Mater Children's, Brisbane. This will be arranged by both cardiologist and then NETS WA team. If there is a simple TGA diagnosed antenatally, then most likely it will be arranged that the mother will go to Melbourne prior to delivery.

POST-OP CONSIDERATIONS AFTER D/C FROM PICU

- The hospital mortality for ASO performed at RCH, Melbourne from 1985-1997 was 0.9% for TGA/IVS and 4.1% for TGA/VSD.
- Following discharge from PICU, complications are rare.
- Late death after newborn TGA has been rare with reports documenting survival at 10 and 15 years as 92-93% and 86% respectively.
- Long term morbidity is due to arrhythmias, valve dysfunction, myocardial ischaemia and neurodevelopmental abnormalities. There are reported incidences of 4.4% AV node dysfunction, 1.7% heart block requiring pacemaker (all had VSD) and 5% SVT at late follow-up. Long term outcome data has shown 3-7% have stenosis or occlusion of the coronary arteries. Current data available suggests that newborn ASO for TGA results in an extremely low risk of long-term neurodevelopmental impairment.
- 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.

National Standards – 1- Care provided by the clinical workforce is guided by current best practice

Legislation - xxxxx

Related Policies - Nil

Other related documents –

RESPONSIBILITY

Policy Sponsor	Neonatology Clinical Care Unit
Initial Endorsement	
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