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Part of the Sydney Children's Hospitals Network

TRANSPOSITION OF THE GREAT ARTERIES WITH INTACT SEPTUM - NETS

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- The neonatologist ± cardiologist at the preferred receiving hospital should be involved in the initial conference call, to assist in making the presumptive diagnosis and discussing management

CHANGE SUMMARY

- Document due for mandatory review

READ ACKNOWLEDGEMENT

- All NETS clinical staff are to read and acknowledge they understand the contents of this guideline

Approved by:	SCHN Policy. Procedure and Guideline Committee	NETS Executive
Date Effective:	1st October 2021	**Review Period**: 3 years
Team Leader:	Staff Specialist	**Area/Dept**: NETS

Rationale

- Transposition of the Great Arteries (TGA) with an intact ventricular septum may be rapidly fatal as the systemic and pulmonary circulations are almost completely independent with only restricted flow across the foramen ovale
- Prostaglandin E1 (PGE_1) (Alprostadil) infusion may not be effective in relieving cyanosis, even though the ductus arteriosus is widely open. The flow of oxygenated blood across the open ductus to the systemic circulation may be insufficient to relieve severe hypoxia and prevent marked metabolic acidosis
- Such neonates may require urgent balloon atrial septostomy (BAS) to survive and this requires a rapid response, 'stabilisation' and transportation
- Although the NETS team must make the retrieval process safe, failure to manage the timeliness of the process; from recognition to surgical intervention may result in a poor outcome

Diagnosis

- Typically there is marked cyanosis in a neonate on the first day of life with initially minimal respiratory distress
- There is often no murmur. The second heart sound is normally split
- Intact septum – no mixing
  - PaO_2 is very low (< 35mmHg) or saturations < 75% and $PaCO_2$ normal or elevated
  - Elevated $PaCO_2$ may indicate respiratory pathology but may also be consistent with a complete parallel circulation
- Ventricular or atrial mixing
  - PaO_2 not so low (35 – 65 mmHg) or saturations 75-90% and $PaCO_2$ normal
- Simultaneous oximetry on the right arm and a leg may demonstrate reverse differential cyanosis (higher saturations beyond the ductus arteriosus)
- The CXR may be normal or show increased pulmonary vascular markings. The classic textbook CXR reveals an 'egg on a string' cardiac silhouette with a narrow mediastinum. However if normal in appearance, in the presence of deep, fixed cyanosis still strongly suggests the diagnosis of TGA[1]
- The ECG is usually normal for age (not an essential investigation and should not delay the process)[1]

Response Where Diagnosis Suspected

- The neonatologist ± cardiologist at the preferred referral hospital should be involved in the initial conference call, to assist in making the presumptive diagnosis and discussing management [2]
- Check that Vitamin K has been given. Important prior to any invasive surgical intervention
- PGE_1(Alprostadil) to open or maintain patency of the ductus arteriosus:
  - Start at 10 nanogram/kg/min. If no response, an increase to 50 nanogram/kg/min may be required; which will likely require ventilation to manage apnoea
  - Intubation and ventilation may be clinically required before the NETS team arrives
- Plan for 'rapid sequence retrieval' for Leg One and Two; to facilitate prompt cardiac assessment and intervention at admission
- Clinical Coordination should complete the ACC tasking request as Priority 1 and requesting Medevac flight category

Leg One

- Leg 1 should be as efficient as possible; including urgent team activation, prompt departure of the NETS team by most timely transport vehicle type for the distance using Priority 1
- Clarify whether a team conference call is required or at team's discretion
- En route to the baby, NETS team to plan an approach to the problem on arrival at the bedside
- Clinical Coordination should book a flight as Medevac cateogory
- The team should confirm Priority & Category on first contact with the aircrew and ask them to facilitate rapid sequence retrieval. For fixed wing and any helicopter landing requiring ground transport, ensure prompt transfer to a road ambulance on landing. Consider the team travelling to the patient separately to the equipment if there are delays
- Pre-load a PGE_1 (Alprostadil) infusion into a syringe driver (unless the referring hospital is known to have started PGE_1 already
- Load any other infusions likely to be used eg. Maintenance (see below), adrenaline and a syringe ready for loading with morphine

Assessment & Rapid Sequence Stabilisation

- Resuscitate the baby – ABC[3]. If shocked, consider and treat infection where the diagnosis unclear
- Check vital signs, including pre/post ductal saturations and BSL

- Look for additional congenital abnormalities that may affect the neonate's condition. E.g. cleft palate & airway abnormalities, gastro-intestinal and neurological abnormalities
- Consider non-cardiac causes of respiratory distress and persistent pulmonary hypertension of the newborn (PPHN); particularly if there is hypercapnoea:
  - Primary PPHN
  - Meconium aspiration syndrome
  - Severe respiratory distress syndrome
  - Sepsis with pneumonia
  - Severe perinatal hypoxia
- These conditions may co-exist with TGA or be the differential diagnosis
- The clinical distinction between persistent pulmonary hypertension of the newborn (PPHN) and a congenital, cyanotic cardiac malformation such as TGA may be difficult to elucidate without an echocardiogram. A history of perinatal fetal distress is common with PPHN. If in doubt, treat as for PPHN
- Cyanosis in TGA is 'fixed' (unchanging despite oxygen) while in PPHN there may be some variability; with the infant also handling poorly. However, the converse can be true
- The presence of respiratory distress is more a feature of lung pathology but can accompany cyanotic congenital heart disease
- Place the oxygen saturation sensor pre-ductally
- Commence PGE_1 infusion (see above)
- There may or may not be an obvious increase in oxygen saturation following commencement of PGE_1
- Prepare for support of the blood pressure with low dose adrenaline - 0.05 microgram/kg/min

Pre-Departure

- If the response to PGE_1 is minimal and the saturations remain critically low – do only the essentials before departure. Do not be delayed by waiting for samples, x-rays, paperwork. In most cases, destination hospitals can see imaging remotely and carrying copies is unnecessary. Conference en route
- No maintenance fluids are required. PGE_1 can be loaded in 10% glucose with an infusion of 50 nanogram/kg/min giving 5 mL/hr (40-60 mL/kg/day) and sufficient to maintain BSL. Treat hypoglycaemia with bolus 10% glucose
- For mechanically ventilated patients given morphine premedication for intubation. A morphine infusion is not required if the transport will be completed within 4 hours. For patients intubated without premedication, morphine a10-20 microgram/kg/hour may be required for analgesia. Don't let this delay departure
- There would only very rarely be a need for placement of central or arterial lines or further x-rays at the referring hospital. Try to leave the umbilicus untouched for use by the cardiologist for septostomy

- Provide the family with information about the likely diagnosis and urgency of cardiac assessment and intervention. Use appropriate local resources (e.g. consultant paediatrician) to make sure the parents are fully informed

Leg Two – En route to the Tertiary Centre

- Plan for an expedited transport that involves as little delay as possible. Priority 1 / Medevac flight category is appropriate
- Use iStat as required for en route blood gases
- In a pressurised aircraft ask for a sea level cabin as baby with suspected TGA may have PPHN and suffer at hypoxic altitudes. In an unpressurised aircraft (helicopter), request LSALT (Lowest Safe ALTitude)

Educational Notes

Transposition of the great arteries (TGA)

- The most common cyanotic congenital cardiac malformation presenting on day 1 of life
- Often not detected antenatally; making post-natal retrieval common (8 per year)[4]
- The aorta arises from the right ventricle and the pulmonary artery from the left ventricle[1]
- Systemic venous blood returns to the right atrium, enters the right ventricle and exits through the aorta
- Oxygenated pulmonary venous blood returns from the lungs, enters the left atrium and the left ventricle and then returns to the pulmonary arteries and the lungs. Atrial communication facilitates mixing of the deoxygenated and oxygenated blood and improves the saturation
- Without some communication between the systemic and pulmonary circulations, there will be severe hypoxaemia
- A communication between the left and right sides of the heart; such as a patent foramen ovale, a ventricular septal defect (VSD), or a patent ductus arteriosus (PDA), singly or in combination, may provide for adequate mixing between the circulations
- The foramen ovale and ductus arteriosus, both normally open in the fetus, usually close soon after birth
- In a subset of neonates with TGA the foramen ovale becomes restrictive very soon after birth. A patent ductus arteriosus can facilitate flow across the foramen ovale and facilitate some rise in saturation. This is how the PGE_1 works to improve saturations in this condition

- If there is no inter-atrial communication even a widely patent ductus arteriosus will not facilitate mixing between the two circulations. Administration of PGE_1 therefore may not improve systemic oxygenation in neonates with suspected TGA in the first hours of life
- The typical neonate with TGA with a restrictive foramen ovale and no VSD presents within a few hours of birth with severe cyanosis; possibly with tachypnoea which is initially minimal but may worsen as metabolic acidosis develops as an attempt to compensate
- TGA may also co-exist with other anomalies such as VSD, PDA, pulmonary valve stenosis, hypoplastic right ventricle and coarctation of the aorta. In such cases, the presenting signs will differ from those described above and the presentation may occur later
- A balloon atrial septostomy establishes an inter-atrial communication to permit adequate mixing between the two circulations and relief of hypoxaemia. This is a temporising procedure which aims to increase saturations to the 80's, correct acidaemia and allow the baby to recover prior to definitive surgery
- Corrective surgery for TGA is an arterial switch operation
- Remember that apnoea is more likely with cyanotic heart disease and PGE_1 at doses greater than 20 nanogram/kg/min[5]

References

1. Jordan S.C., Scott, O. Heart Disease in Paediatrics; 1973; Butterworths and Co Ltd; Great Britain.
2. Shivananda S. Kirsh J. Whyte HE. Muthalally K. McNamara PJ Accuracy of clinical diagnosis and decision to commence intravenous prostaglandin E1 in neonates presenting with hypoxemia in a transport setting. Journal of Critical Care. 25(1):174.e1-9, 2010 Mar.
3. ILCOR Guidelines 2010
4. Woods T, Browning Carmo K, Wall M, Berry A Transporting Newborns with transposition of the great arteries Jnl of Pædiatrics and Child Health 49 (2013) E68–E73
5. Browning Carmo K, Barr P, West M, Hopper NW, White JP, Badawi N Transporting newborn Infants with suspected duct dependant congenital heart disease on low dose prostaglandin E1 without routine mechanical ventilation ADC 2007 (92)