Guideline for Care of babies with Antenatally Diagnosed Transposition of Great Arteries and Intact ventricular septum/ small ventricular septal defect (TGA-IVS)

Applies to BWH, BCH and KIDS-NTS

Version:	*2*
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Purpose of the guideline:	To provide a pathway process for the management of babies with antenatally diagnosed TGA with intact ventricular septum/ small VSD who may require an Atrial Septostomy
Who should use the guideline?	*Any member of staff involved in the care of a baby with antenatally diagnosed TGA with intact ventricular septum/ small VSD who may require an Atrial Septostomy.*
How was the guideline developed?	*With input from Cardiology, Cardiothoracic surgery, KIDS-NTS, PICU, NICU and Obstetric/maternity teams.*
How will the guideline be monitored?	Via Datix and feedback from all teams involved with the care of TGA babies who require Atrial Septostomy
Approved by:	Neonatal Clinical Practice Meeting
Date Approved:	
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Content:

- Transposition of the Great Arteries = TGA
- Balloon Septostomy = BS
- BWC = Birmingham Women's and Children's Foundation Trust (Mindelsohn Way)
- BCH = Birmingham Children's Hospital site (Steelhouse Lane) BWH
- KIDS/NTS Transport Team
- BC PICU = Birmingham Children's Paediatric Intensive Care Unit
- BW NICU = Birmingham Women's Neonatal Intensive Care Unit
- DS = Delivery Suite

2. Introduction and background:

- Transposition of the great arteries is a duct dependent congenital heart defect where the outflow tracts (aorta and pulmonary artery) are reversed, creating parallel circulations. The ventricular septum can be intact, or there can be an associated ventricular septal defect(s) which can be small or large.

- Transposition of the great arteries can also be associated with other heart lesions, but this guideline relates only to Transposition of the great arteries with an intact ventricular septum or with small ventricular septal defect(s).

- The number of cases with transposition of the great arteries that are antenatally diagnosed has increased to over 70% in some regions of the United Kingdom (www.nicor.co.uk)[1, 2].

- Oxygenation postnatally is reliant on mixing of oxygenated and deoxygenated blood via an inter-atrial communication (PFO/ASD). A patent ductus arteriosus (PDA) can improve mixing at the atrial level. Mixing may also take place at ventricular level if there is a large ventricular septal defect, but this is not always the case.

- Pulmonary hypertension (increased pulmonary vascular resistance) after birth may also reduce mixing at atrial level, reducing oxygenation.

- Every baby with transposition of the great arteries initially requires a continuous infusion of prostaglandin to maintain patency of the ductus arteriosus.

- If there is not adequate mixing due to a restrictive atrial communication postnatally, then a **time critical balloon atrial septostomy (BAS)** is required.

- **Antenatally, the shunting across the atrial septum cannot be predicted with certainty**, **therefore one should be prepared for a time critical transfer to BCH for septostomy[3, 4].**

- This guideline and SOP have been developed to provide the various teams involved in the care of a baby with TGA with intact ventricular septum/ small VSD a clear pathway to coordinate the care from antenatal phase to resuscitation and stabilisation at birth and transfer to theatre/PICU for likely atrial septostomy.

3. Content:

3.1 Antenatal care:

- All babies with an antenatal diagnosis of TGA should be delivered at BWH. There is a small population who may be diagnosed postnatally.

- These mothers will have regular scans and counselling provided by Fetal Cardiology, Fetal Medicine and neonatal teams. **All prenatally diagnosed TGA with intact ventricular septum/ small VSD will be managed on this pathway**.

- The parents will be fully informed at all points of the pathway so they can be engaged and aware of the plans and possible outcomes. The information would include the split site working between BWH and BCH and the urgency of transfer. It should be made clear to the mother during counselling that some babies with TGA are extremely sick from birth and do poorly.

- A planned delivery (either IOL or elective section) may be offered according to obstetric indications and the patient's wishes. The obstetric management of labour is not dictated by the fetal diagnosis. The risks and benefits of elective CS versus induction of labour should be discussed including any logistical aspects of care such as transportation, with informed parental choice to be respected. **If caesarean section delivery is planned then this should be prioritised on the operating list, as it impacts the patient flow at KIDS-NTS, PICU and theatres. The caesarean section should be first on the obstetric list, as hybrid theatre or catheter laboratory activity may be delayed until it is known that the baby does not need an emergency septostomy. It is important to recognise that this may not always be possible.**

- The Maternity/Fetal Medicine clinical team (or if scheduled for caesarean section, the theatre booking co-ordinator) will inform the Neonatal Matrons/ HoN /Clinical Lead of all upcoming cot dependant pregnancies, **with Name, Date of Birth, Hospital Number, Fetal diagnosis and Expected Date of Delivery**, and this will be communicated by the Neonatal Team via phone to the BCH cardiac, Intensive care and KIDS-NTS teams as early as possible.

3.2 Delivery/Induction:

- At admission to hospital the NICU Nurse in charge/ Matron/ Neonatal Consultant will be informed that the woman is on site, by the obstetrician or Midwife in charge or Delivery Suite Matron.

- This information should be shared with the rest of the Neonatal team as soon as possible and highlighted at the daily Huddle (every day until the baby is born) to allocate teams to attend the potential delivery.

- **NICU, PICU bed and Cardiology consultant on call should be informed before induction (membranes broken)/ caesarean section delivery by a conference call via KIDS-NTS**. Progress should be regularly communicated to the teams (obstetric, cardiology, NICU and KIDS) when the delivery is near, for logistical planning.

- At the time of labour (spontaneous or induction) or Caesarean section there will be communication with NICU Nurse in charge/Consultant who will communicate with KIDS and NTS consultants via the KIDS-NTS number to decide which team is best able to do the time critical transfer. The KIDS/ NTS consultant will aim to be present with the team (to maintain situational awareness) at the time of delivery for the time critical transfer.

- When delivery is imminent, KIDS-NTS team will be informed as a matter of urgency. A **conference call will be arranged by the Neonatal Consultant via the KIDS-NTS number 03002001100 which should include KIDS and NTS consultant, Cardiologist on call, and PICU consultant. Cardiologist on call will inform the Interventional Cardiologist. The interventional cardiologist will ensure that theatres/ catheter laboratory (Angio theatre) team are aware. They may designate a junior member of the cardiac team to do this. The neonatal team will coordinate with the maternity team (midwife co-ordinator/ Obstetric consultant). Maternity team can also be involved in the conference call.**

- The attending team at delivery must include neonatal consultant, Tier 2 doctor/ANNP, tier 1 doctor and a senior neonatal nurse for an anticipated need for urgent septostomy. The KIDS-NTS team should also aim be present on site.

- There would be always a risk that both teams (KIDS/ NTS) are out on transport and the delivery should be postponed (if it is possible to do so).

3.3 Neonatal care:

DELIVERY ROOM/THEATRE:

- BW NICU team will **prepare in advance to attend the delivery**.
    - **Identify if intact ventricular septum or small VSD.**

        - **Prepare in advance**
            - 2 cannulation sets
            - Double/triple lumen UVC (no X-ray required for time critical transfer)
            - Consideration of intra-osseous access in the challenging venous access situation
            - On **estimated fetal weight** following drugs and infusions to be made
                - Prostaglandin infusion
                - Morphine bolus and infusion
                - Rocuronium bolus and infusion
                - 10% glucose for maintenance
                - Adrenaline infusion (for peripheral and central administration)
                - Vitamin K
                - First line antibiotics

        - **Summary sheet of the baby details**
            - Antenatal findings
            - Gestation
            - Estimated weight
            - Parent details (name, NHS number and contact phone numbers)
            - Baby identification bands 2 with NHS and BW number (to be filled as soon as born and registered)

- BW NICU team Attending teams should be clearly identified, and roles clearly allocated to avoid confusion. A neonatal consultant should be in attendance.

- The NICU team will work collaboratively with the KIDS-NTS team to help with resuscitation and stabilisation.

- The baby will be resuscitated as per neonatal life support (NLS) guidelines.

    - **Preductal SpO2 75-85%** are **acceptable**. Stable babies with normal breathing and **SpO2 >75% do not require immediate intubation** and can be transferred to PICU on prostaglandin infusion for on-going management.

    - **If pre-ductal SpO2 <70% after the first 5 minutes**, despite increasing oxygen to 100%

- **Airway**: Intubate (ideally with rocuronium and morphine bolus given via a line)

- **Breathing**: Ensure gentle ventilation and avoid hyperventilation (as it can increase pulmonary blood flow). Do not do an X-Ray to confirm lines/ ET position as that can be confirmed in PICU/ Theatres once time critical transfer done.

- **Circulation**:
- Cannulate with 2 intravenous access (2 cannulas/ double/ triple lumen UVC)
- Start Prostaglandin infusion (5-50 ng/kg/min)
- Adrenaline infusion (0.01- 0.2 micrograms/kg/min)
- Antibiotics (**not obligatory if no risk factors for sepsis**)

- **Disability:**
- Give morphine, rocuronium bolus
- Start Morphine and Rocuronium infusion. If infusions are not ready the baby can be transferred on boluses (rocuronium/ morphine)
- Give Vitamin K
- Get baby labels and write a page summary of care
- **KIDS-NTS to do a time critical transfer**
- BWH and KIDS-NTS team to update the parents about the high-risk transfer

- **To note, if baby is hypoxic despite above interventions, no time should be wasted in getting more clinical information, as <u>the only intervention that will improve the clinical condition is a septostomy</u>**

- **Transfer Cat 1 (on blue lights) to PICU for assessment or diversion to the Catheter Laboratory (Angio theatre).**

- **The KIDS team or neonatal consultant should contact the cardiology consultant via a conference call. The cardiologist will inform the interventional cardiology consultant on call with an update**.

- **If the baby is clinically unstable, significantly desaturated or acidotic, the catheter laboratory team should be mobilised immediately and baby transferred directly to the catheter laboratory.**

<u>3.4 PICU/ Catheter Laboratory guidance</u>

- All babies with TGA/IVS will be considered as needing septostomy on an urgent basis.

- Once a decision is made for BAS, the baby should be added to emergency list via electronic patient record ~~(Taurus)~~. All middle grade doctors should be given necessary training for the same.

- The cardiologist will contact the cardiac anaesthetist on call. The on-call registrar will be expected to call the theatre co-ordinator, Angio radiographer and cardiac physiologist if required by the interventional cardiologist. If this is out of hours and if non-invasive cardiologist is on call, then they will be expected to help with some of the process, and it should not be responsibility of the interventional cardiologist alone.

- Location of transfer – This will depend on the clinical condition of the baby. The final decision of where the baby should be transferred to will depend on the discussion between KIDS/NTS, and the on-call cardiology team ~~on shop floor~~ and logistics of the day.

- Procedural consent should be done in person or over the phone depending on urgency and availability of mother/ father of the baby. The standard trust guidelines for consent will apply.

- Once transferred a rapid ECHO will be performed to confirm diagnosis and assess the atrial septum.

- If indicated, Balloon atrial septostomy will be performed under GA by the interventional cardiologist. If a non-interventional cardiologist is on call, then they will provide support for the Echo guidance with the on-call registrar if available. In a stable child a cardiology trainee will be allowed to do the septostomy but under the guidance of an interventional cardiologist who will carry the responsibility for the procedure.

- Balloon atrial septostomy is a relatively standard procedure and will be performed either in cardiac catheter laboratory or hybrid theatre and rare instances the IR suite. X ray equipment is likely to be needed or at least needs to be available due to the nature of the current equipment used for BAS.

- BAS is a standard procedure and please refer to BAS document for details regarding the technical details.

- There are rare times when ECLS may be considered instead of doing BAS. This will be a case-to-case decision depending upon clinical circumstances. This will require a discussion between the ECLS and Cardiology teams. ECLS or ECLS backup could be considered if there is a very high lactate (>10) from the outset and failure to improve, significantly impaired LV function at the outset, BAS attempted but cardiologist unable to cross the septum despite reasonable attempts and persistent significant hypoxia.

- In any other situation, including ECPR, the final decision will be with the team

managing as it is impossible to cover all possible scenarios, but it is anticipated this will be a rare situation.

3.4 Post procedure care/plan:

- Following the Septostomy the baby will be transferred to BC PICU, as above.

- If a BC PICU bed is not available, then there will be a discussion regarding the possible transport of the baby back to BW NICU. This is dependent upon the baby's condition and NICU capacity.

- Prostin can be left at 5 nanograms/kg/min after the BAS for 12 to 24 hours and then consider stopping depending on the clinical status of the baby. In some babies despite of adequate inter-atrial communication Prostin is needed to maintain SAO2 > 75%.

- In a stable child with SAO2 > 75% and Lactate < 3 consider weaning ventilation sooner.

References:

1. https://www.nicor.org.uk

2. Bonnet D, Coltri A, Butera G, Fermont L, Le Bidois J, Kachaner J, Sidi D. Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality. Circulation. 1999 Feb 23;99(7):916-8. doi: 10.1161/01.cir.99.7.916. PMID: 10027815.

3. Stockley, EL, Singh, A, Desai, T & Ewer, A 2019, 'Can fetal echocardiograms reliably predict the need for urgent balloon atrial septostomy in transposition of the great arteries?', Archives of Disease in Childhood, vol. 104, no. 11, pp. 1114-1116. https://doi.org/10.1136/archdischild-2019-317867.

4. Gottschalk, I., Walter, A., Menzel, T. *et al.* D-Transposition of the great arteries with restrictive foramen ovale in the fetus: the dilemma of predicting the need for postnatal urgent balloon atrial septostomy. *Arch Gynecol Obstet* **309**, 1353–1367 (2024). https://doi.org/10.1007/s00404-023-06997-8