Title:	Transposition of the Great Arteries without VSD
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Target Audience	**CM Neonatal Units and potentially other units referring babies into the CM Network**
Reference Number:	**CM-GL-07**
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Version	**Final**

Date	Version	Author	Notes
9/10/21	v0.1	NS	This supersedes the previous TGA guideline which was TGA with intact VSD. Cardiac Network up-dating CHD guideline but TGA to continue to be overseen by the Neonatal ODN.

Transposition of the Great Arteries without VSD

Background
Babies with transposition of the great arteries without a ventricular septal defect (TGA without VSD) can develop life-threatening profound hypoxaemia quickly after birth. Mixing of oxygenated and deoxygenated is minimal with a restrictive atrial communication even in the presence of a widely patent ductus arteriosus. Urgent treatment with atrial septostomy is required and can be life-saving. In other babies with TGA without VSD there is a non-restrictive foramen ovale which allows adequate mixing at atrial level to sustain safe levels of systemic oxygenation.

Delay in providing treatment for babies with a restrictive foramen ovale increases the risk of death.
It is not possible to be certain from antenatal imaging to determine with any accuracy which fetuses with TGA without VSD will have a restrictive or non-restrictive foramen ovale after birth. For this reason, all babies with an antenatal diagnosis of TGA without VSD must be transferred to Alder Hey Hospital as quickly as possible after birth.

<u>This transfer is not to be delayed by a period of assessment or by waiting for impact of any other treatments that are intended to improve oxygenation or acid base status. Septostomy is the only effective treatment for the acute profound hypoxaemia seen with TGA without VSD.</u>

The actions described below are a set of recommendations for best practice, agreed locally. Although the research evidence base for these recommendations is weak, this document represents a framework of practice with which to manage such babies.

Actions
Please refer to TGA without VSD pathway document and complete checklist.

1. <u>Antenatal management</u>
   - Joint counselling of parents should be performed by fetal medicine, cardiology and neonatal teams;
   - Diagnosis and implications should be discussed in detail with parents supported by a parent information leaflet;
   - Consider parallel planning with involvement of hospice teams if appropriate;
   - The timing, mode and place of delivery should be agreed with the parents and the discussion and decision should be clearly documented;
   - Although the preferred pathway should be elective CS at LWH, informed parental choice should be respected;
   - Delivery might be elective (induction or CS) or following spontaneous labour;
   - Schedule CS to be first case on the morning list;
   - Enter details of planned delivery in Fetal Medicine and Neonatal Outlook Diary.

2. Pre-delivery management

a) If the parents opt for CS/induction:
- Arrange delivery at LWH at around 39-40 weeks' gestation. Elective CS should be arranged for a Tuesday/Wednesday if possible and induction should not take place on a Sunday;
- 24h pre-elective CS delivery/induction: Elective CS consultant or DS Shift leader (as appropriate) to inform neonatologist (bleep 333 and/or at daily DS huddle); Neonatologist to inform Connect North-West, AH Cardiology and PICU to confirm decision to proceed with delivery via conference call;

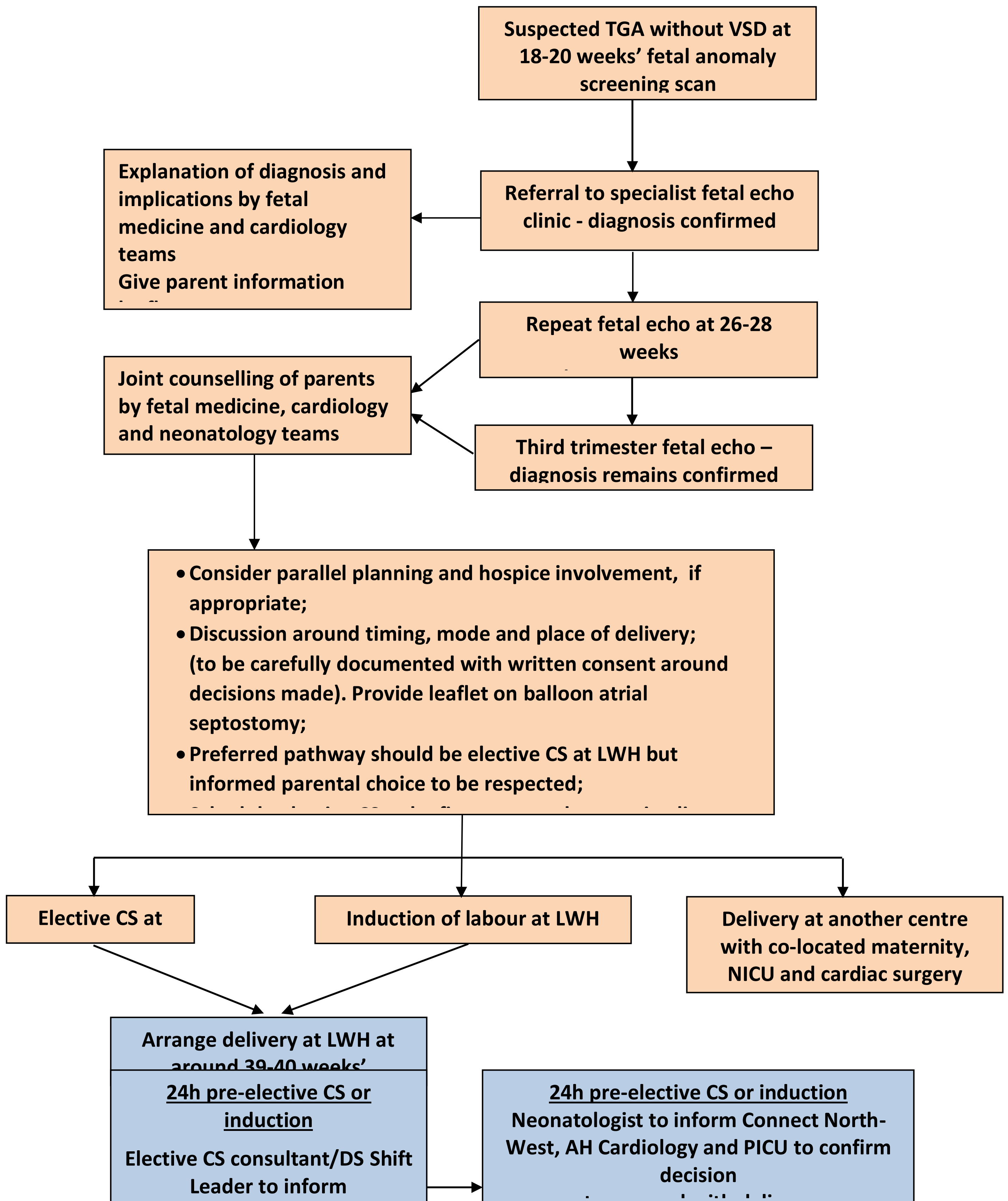
b) If spontaneous labour:
DS Shift leader to inform neonatologist as early as possible and neonatologist to alert Connect North-West, AH Cardiology and PICU teams via conference call.

3. Delivery room management
- 08:30 on day of delivery: neonatologist to arrange conference call with Connect North-West/AH Cardiology/PICU;
- Prepare prostin infusion before delivery;
- Prepare pre-med intubation drugs (fentanyl/suxamethonium);
- Ensure equipment available for UVC insertion;
- Ensure person inserting UVC/assistant are scrubbed and prepared;
- Contact radiographer and Connect North-West close to time of delivery.

4. Immediate neonatal care after delivery
- Insert and secure double-lumen UVC in delivery room;
- Start prostin at 10 ng/kg/min;
- Give supplemental oxygen if SpO2 < 75%;
- Give fentanyl/suxamethonium;
- Routinely perform tracheal intubation in delivery room;
- Inform Connect North-West of delivery;
- Transfer to NICU;
- Start morphine infusion at 10 mcg/kg/hour;
- Perform capillary blood gas within 30 minutes of birth;
- Perform chest x-ray;
- Liaise with Connect North-West about transfer to Alder Hey;
- Inform Cardiology and PICU when baby leaves LWH including update about condition/gas.

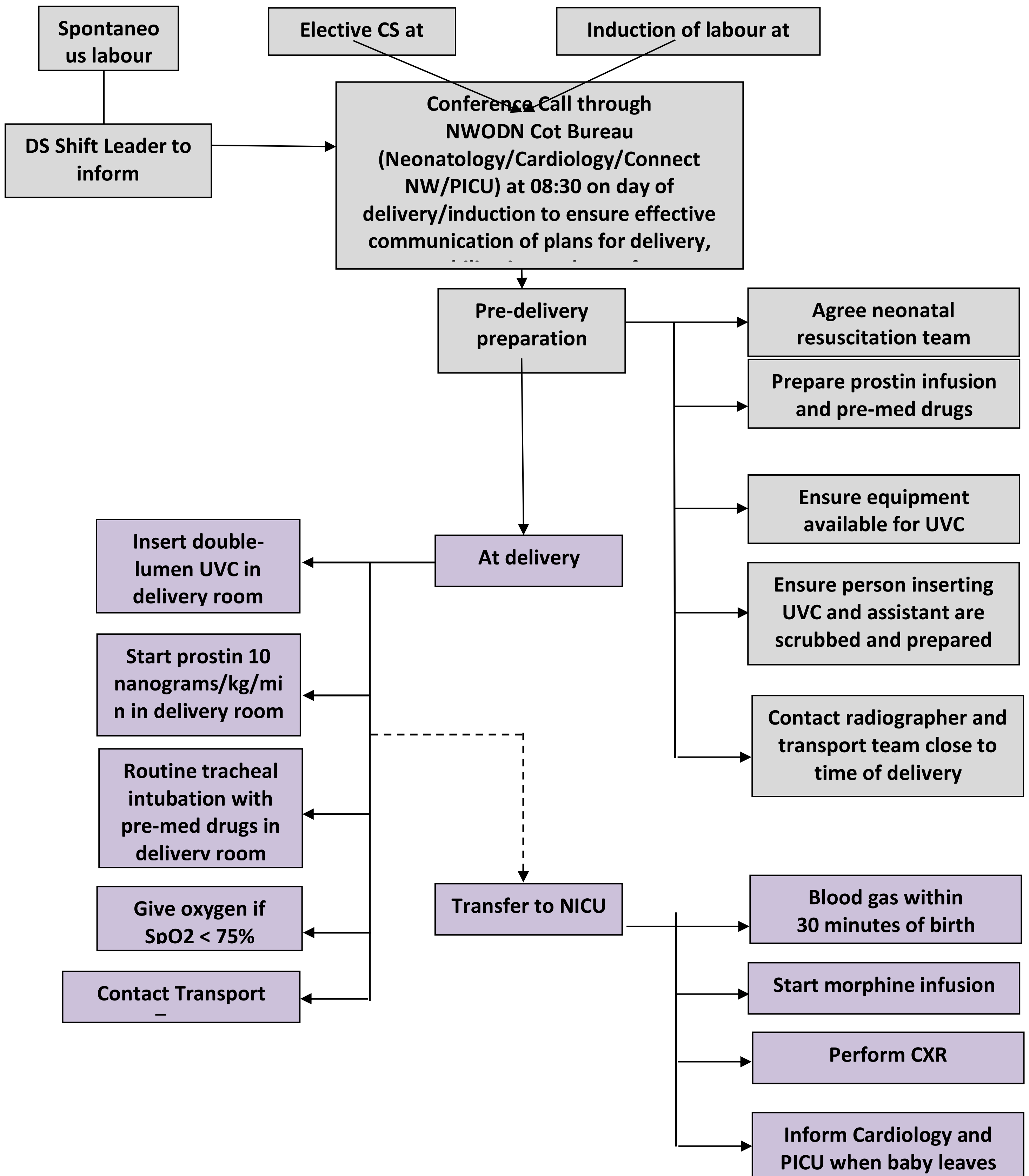
TGA without VSD pathway

(Part 1 – Antenatal and pre-delivery management)



TGA without VSD pathway

(Part 2 – Delivery room management and immediate neonatal care)



TGA without VSD pathway (part 1):

FETAL MEDICINE UNIT CHECKLIST – print out and file in maternal notes (and into baby notes after delivery)

Mother's Name____________________________ LWH number________________________

Baby's Name____________________________ LWH number________________________

ANTENATAL MANGEMENT			
	Date	Print name	Initials
Fetal echo (1) performed to confirm diagnosis			
Information leaflet given			
Parents counselled about cardiac risks and informed of initial management plan			
Fetal echo (2) performed to confirm diagnosis			
Antenatal booking (if required)			
Joint counselling of parents with FM, cardiology, neonates			
Parents offered hospice involvement, if appropriate			
Discussion around timing, mode and place of delivery and decision documented			
Date of induction			
Date/time of CS			
Enter details of planned delivery in Fetal Medicine and Neonatal Outlook Diary			

TGA without VSD pathway (part 2):

NEONATAL UNIT CHECKLIST – print out and file in baby notes

Mother's Name____________________________ LWH number________________________

Baby's Name____________________________ LWH number________________________

PRE-DELIVERY MANAGEMENT			
	Date/time	**Print name**	**Initials**
24h pre-CS or induction (or as early as possible after spontaneous labour): Elective CS consultant/DS shift leader to inform neonatologist (bleep 333 or at joint DS huddle)	**(When neonatal team first contacted by maternity staff)**		
24h pre-CS or induction (or as early as possible after spontaneous labour): Neonatologist to inform Connect NW/AH Cardiology/PICU			

IMMEDIATE NEONATAL CARE AFTER DELIVERY			
	Date/time	**Print name**	**Initials**
Insert UVC in delivery room			
Give pre-med intubation drugs			
Routine tracheal intubation in delivery room			
Start prostin 10 nanograms/ kg/min in delivery room			
Inform Connect NW of delivery			
Admission to NICU			
Blood gas within 30 minutes of birth			
Start morphine infusion			
Perform CXR			
Inform Cardiology and PICU when baby leaves LWH			
Transferred to AH			

Useful contact numbers

Connect North West/Cot Bureau 0300 330 9299
Alder Hey Consultant Cardiologist 0151 252 5220 (or via AH switchboard 0151 228 4811)
Alder Hey PICU (direct) 0151 252 5241

TGA without VSD prescription chart

Name	Date of birth	W………………………………………

Single Prostin E2

Draw up 500 micrograms of Dinoprostone (Prostin E2) which is 0.5mL of a 1mg/mL solution. Dilute with 10% dextrose to a total volume of 500 mLs. This will give a solution with a final concentration of 1 micrograms/mL. Withdraw 50mLs of this solution from the bag into a syringe to use for the infusion

Run at 10 nanograms/kg/min (range is 10-to 100 nanograms/kg/min)

Signed………………………………Printed…………………………Date………………………………

Given by ……………………..…………….. Checked by…………………….………………………

10% Dextrose

Run at 7.5 ml/hour. This is equivalent to total daily fluids of 60ml/kg/day for a 3Kg baby

Signed………………………………Printed…………………………Date………………………………

Given by ……………………..…………….. Checked by…………………….………………………

Date	Time	Working weight 3 kg	Dose 2mg / kg	Calculated Dose 6 mg	Route IV	Date	Time
Approved Medication Suxamethonium			Special Instructions fast intravenous bolus over 5 - 10 seconds		Pharmacy	Given By	
Signature			Print Name			Checked by	

Date	Time	Working weight 3 kg	Dose 5 micrograms/kg	Calculated Dose 15 micrograms	Route IV	Date	Time
Approved Medication Fentanyl			Special Instructions Slow IV bolus over 3-5 minutes.		Pharmacy	Given By	
Signature			Print Name			Checked by	

Date	Time	Working weight 3 kg	Dose 1mg	Route IM	Date	Time
Approved Medicine VITAMIN K (Konakion MM Paediatric)			Special Instructions	Pharmacy	Given By	
Signature			Print Name		Checked by	

Morphine Maintenance - PRE-FILLED SYRINGE

Use prefilled morphine infusion 20ml syringe of DOUBLE strength (100 micrograms/ml).
Infuse at 10 – 30 micrograms/kg/hr

Signed……………………………….Printed…………………………Date…………………………….

Given by ……………………………………… Checked by……………………..……………………….