Guideline for the perinatal management of the fetus with antenatal diagnosed cardiac heart disease (CHD).

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The North West Neonatal Network (NWNODN) consists of 3 locality neonatal networks, Cheshire and Merseyside (CM) Lancashire and South Cumbria (LSC) and Greater Manchester (GM). This
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- North West and North Wales Paediatric Transport Service
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- S Vause
Guideline for the perinatal management of the foetus antenatally diagnosed with cardiac disease (CHD)

Introduction

This document on the perinatal management of the foetus with antenatally diagnosed CHD is intended to provide guidelines for selection of delivery site for the foetus with antenatally diagnosed congenital heart disease (CHD). The overriding aim is to deliver a quality foetal cardiac service across the North West Operational delivery Network (NWNODN); to improve early outcome through the provision of appropriate and prompt early neonatal care with transportation to the appropriate cardiac centre if necessary.

The North West is a large region encompassing many obstetric and neonatal units, therefore care prenatally for the foetus with CHD and the mother will involve multiple professionals, in often geographically remote locations. This pathway aims to structure consistent delivery planning and improve communication between relevant professionals.

Fetal cardiology expertise is currently delivered at Liverpool Women’s Hospital (LWH) and St Mary’s Hospital (SMH) in Manchester. On both sites this involves a multidisciplinary approach of foetal cardiologists, foetal medicine specialists, neonatologists, specialist midwives and sonographers. Cardiac liaison nurses at Alder Hey Children’s and Royal Manchester Children’s Hospitals also provide information, support and guidance in the antenatal period to prepare families prior to delivery.

The vast majority of congenital heart disease seen in the foetus represents a spectrum and care must be individualised. Delivery planning will take into account many factors including the particular cardiac condition and physiology as assessed on foetal echocardiogram, parental preference, family circumstances and local neonatal care provision. The foetal cardiologist or foetal medicine specialist will lead perinatal planning in collaboration with the obstetrician, neonatologist and family. An effort is made to deliver care locally where possible.

For the majority of families an antenatal cardiac diagnosis is made at around 20 week’s gestation after the routine anomaly scan. Following initial diagnosis usually a detailed scan to check the extracardiac anatomy will be offered and perinatal planning should begin early with the family. Final plans should be confirmed typically later in gestation between 28 and 34 weeks and documented clearly in the maternity notes. Local obstetric care is essential and should continue throughout pregnancy. Communication with all professionals involved is vital.

Cardiac surgery in the region is performed at Alder Hey Children’s Hospital (AH) and children’s cardiology services are also delivered at Royal Manchester Children’s Hospital (RMCH). The network provides many peripheral outreach clinics so cardiology follow-up is available closer to home.
Notes

Following delivery

- **Please read the fetal cardiology reports and follow the documented management plan**
  - Particular reference should be made to whether iv Dinoprostone (Prostin) should be commenced.
  - Initial dose should be 5 nanograms/kg/minute in antenatally diagnosed patients if there are no clinical concerns. Higher doses maybe needed in those with haemodynamic compromise.
- Contact the relevant cardiology team to inform them of the baby’s birth and arrange appropriate review
  - A **faxed or digital referral** should be sent ahead of all referred patients – this should contain: referring paediatric consultant; patient name, address, DOB and NHS number; mother’s name; patient’s GP details
  - Please send a copy of the foetal cardiology report along with the baby.
- If transport is required liaise early with the relevant team

Balloon Atrial Septostomy (BAS)

- All patients likely to require to BAS i.e. transposition of the great arteries with intact ventricular septum are currently to be delivered at LWH. Please refer to the Liverpool Women’s Guideline for the Management of the Antenatal diagnosis of Transposition of The Great arteries ( Appendix A)
- Please call Alder Hey cardiology SpR when the mother is admitted for planned delivery so the on-call catheter lab team can be pre-warned.

Non-duct dependent lesions

- Those with non-duct dependent lesions are expected to be well after delivery.
- Any neonate who is unwell should be discussed urgently with the relevant cardiology team.
- Neonates with Atrio-ventricular septal defect and Tetralogy of Fallot should be observed on the special care baby unit for a minimum of 48 hours following delivery. Saturations should be monitored.
- All patients should be discussed with cardiology (and plans should be in place for review) prior to discharge from hospital.
- In the majority of cases where the baby is well and no monitoring is required, discharge and transportation to Alder Hey or Manchester in parents’ car should be possible.
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**Antenatal Referral Table**

**Referral to foetal medicine unit**
Specialist foetal cardiology assessment to be performed within 3 working days.

**Neonate requiring urgent cardiology review to guide management**
- Univentricular heart with duct dependent pulmonary or systemic circulation e.g. hypoplastic left heart syndrome
- Critical aortic stenosis
- Transposition with VSD
- Sustained fetal arrhythmia e.g. complete heart block, supraventricular tachycardia
- Complex cardiac and pulmonary interactions e.g. Absent pulmonary valve, Severe Ebstein’s anomaly, total anomalous pulmonary venous connection
- Biventricular heart suspected duct dependent pulmonary or systemic circulation e.g. Pulmonary atresia or ToF with severe PS, hypoplastic aortic arch, coarctation of the aorta, critical pulmonary stenosis
- Univentricular heart, non-duct dependent e.g. Double inlet left ventricle
- Non-duct dependent lesions e.g. Tetralogy of Fallot well developed PAs

**Liaison between relevant specialist teams**
(foetal medicine, cardiology, neonatology, palliative care, obstetrics and local paediatric team)
**Individualised perinatal management plan** to be agreed with parents and documented in medical and maternity notes

**Delivery at LWH**
*to facilitate urgent septostomy at AH

**Stabilisation and transfer to AH within 6 hours of birth**
if severe hypoxaemia or acidaemia, contact AH team to discuss earlier transfer

**Delivery at LWH/SMH**
*to facilitate early tertiary cardiologist review

**Deliver locally if NICU provision, if not transfer to LWH/SMH**

**Review by/ discussion with cardiologist within 24 hours after birth**
This is a minimum standard, many of these neonates will require earlier cardiac assessment
If urgent surgery (<48 hours) necessary contact AH team to discuss early transfer to surgical centre
If clinical concerns, severe hypoxaemia or acidaemia for stabilisation and urgent transport to RMCH or AH

**Delivery at local unit**

**Postnatal assessment**
Discussion with cardiology team at RMCH/AH prior to discharge

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Important Contacts Details:

The Foetal Cardiology Teams:

Liverpool Women’s Hospital
- Foetal Medicine Unit: 0151 702 4072
- Cardiac Liaison Nursing Team at Alder Hey: 0151 252 5291

- Dr Joyce Lim  Consultant Fetal Cardiologist
- Dr Devender Roberts  Clinical Lead Fetal Medicine
- Dr Umber Agarwal  Consultant in Fetal Medicine

St Mary’s Hospital, Manchester
- Foetal Medicine Unit: 0161 276 6385
- Cardiac Liaison Nursing Team at Royal Manchester Children’s: 0161 701 0664

- Dr Gordon Gladman  Consultant Fetal Cardiologist
- Dr Caroline Jones  Consultant Fetal Cardiologist
- Dr Philip Bullen  Clinical Lead Fetal Medicine
- Dr Koon Loong Chan  Consultant in Fetal Medicine

Cardiac Centre’s: Alder Hey Children’s Hospital Switchboard  0151 228 4811

On call registrar bleep  369
Cardiac secretaries  0151 252 5711
AH Fax referrals  0151 252 5643
Ward K2 (cardiac unit)  0151 252 5418
PICU  0151 252 5242

RMCH Switchboard  0161 276 1234
RMCH Cons Phone  0161 701 7474
Cardiac secretaries  0161 701 2179
RMCH Fax referrals  0161 701 0619
Ward 85 (cardiac unit)  0161 701 8500
PICU  0161 701 8000

Transport Teams
- Connect North West (Neonatal transport team)  0300 330 9299
- NWTS (Paediatric transport team)  08000 848 382
Appendices:

Appendix A:

POLICY FOR THE MANAGEMENT OF ANTENATAL DIAGNOSIS OF TRANSPOSITION OF THE GREAT ARTERIES WITH INTACT VENTRICULAR SEPTAL DEFECT (LWH)

Background

Babies with transposition of the great arteries with an intact ventricular septum (TGA/IVS) can develop life threatening profound hypoxaemia quickly after birth because the intra-atrial connection via the foramen ovale can be restrictive and mixing of oxygenated and deoxygenated blood may be minimal even in the presence of a widely patent ductus arteriosus. For these babies urgent treatment with atrial septostomy is required and can be life saving.

In some babies with TGA/IVS there is a non-restrictive foramen ovale which allows adequate mixing via the foramen ovale 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/IVS will have a restrictive or non-restrictive foramen ovale after birth. For this reason, all babies with an antenatal diagnosis of TGA?IVS must be transferred to Alder hey as quickly as possible after birth. 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 profound hypoxaemia seen with TGA/IVS.

Agreed plans.

If the diagnosis is TGA/IVS with unbalanced ventricles.

Consideration should be given to delivery in a co-located centre such as Guys and St Thomas’s Hospital or Leeds. This is to be discussed in the antenatal period on a case by case basis.

If the diagnosis is TGA/IVS with balanced ventricles.

We will aim to deliver all other women having a foetus with TGA/IVS in Liverpool. They should all be offered delivery by caesarean section with explanation of the risks of normal delivery (potential non-availability of transport etc.).

For women who agree to delivery by planned caesarean section

When the date and time of the delivery is agreed in the antenatal period, the neonatal team, cardiology team and neonatal transport team must be informed.

Communication on the day of admission:

The maternity team will inform the neonatal team when the woman is admitted.
Working together to provide the highest standard of care for babies and families

The Neonatal team will inform the cardiology team.

The cardiology team to liaise with PICU team and confirm to neonatal team that cardiology and PICU beds are available.

Neonatal team to liaise with the Neonatal Transport team (Connect NW) to ensure that they will be available at the agreed time of the CS.

If the transport team are not available due to already being involved in retrieving other babies, the caesarean section will not proceed until they are available.

At delivery

The neonatal team to have Prostin drawn up prior to birth.

An umbilical venous catheter is to be inserted immediately at birth, in the theatre and a Prostin infusion commenced immediately.

A chest x-ray will be performed to confirm safe placement of UVC (If the tip is in an intracardiac position, the UVC should be repositioned. A low lying UVC is safe for short term Prostin administration and can be left in place). The endotracheal tube position will also be assessed on the chest x-ray if the baby has been intubated.

The baby is then to be transferred immediately to Alder Hey.

The transport team will liaise directly with cardiologist on call to determine where the baby is to be taken (cardiology ward, PICU, catheter lab).

Attempts to ‘stabilise’ the baby before transfer are futile, immediate transfer must take priority over all other interventions.

For women who chose normal delivery

Communication on the day of admission:

The maternity team will inform the neonatal team when the woman is admitted.

The Neonatal team will inform the cardiology team.

The cardiology team to liaise with PICU team and confirm to neonatal team that cardiology and PICU beds are available.

The Neonatal Transport team (Connect NW) to be informed when cervix is dilated to 8cm and will then attend.

If the Neonatal Transport team are unavailable and no alternative team can be mobilised by Connect NW, NWTS will be called to perform transfer.

Attempts to ‘stabilise’ the baby before transfer are futile, immediate transfer must take priority over all other interventions.

If there is no means by which the baby can be rapidly transferred to AHCH and the baby is unstable, the cardiologist should consider whether it is possible to do an ultrasound guided septostomy at LWH.
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