

Perinatal planning for Pre-natally diagnosed congenital heart disease

This document acts as a general guide for planning the delivery of prenatally diagnosed cases of congenital heart disease. It should be remembered that perinatal management planning may need to be tailored for individual cases taking account co-morbidities and parental preference with involvement of all appropriate members of the multi-disciplinary team.

In all cases, echo reports and letters from the fetal cardiology team should be available in the hand-held maternity notes, sent to the neonatal/paediatric team in the planned delivery hospital. The reports should contain information regarding the planned location and mode of delivery and also on the anticipated postnatal treatment strategy. On a monthly basis, Dr Abu-Harb will update the paediatric cardiac team at FRH of cases expected to deliver that month.

Deliver in Freeman:

This will be required only rarely (1 case every 2 years) and before this is decided there should be a multi-disciplinary discussion including fetal medicine, neonatology, paediatric cardiology, paediatric cardiac surgery and paediatric cardio-thoracic intensive care. Delivery at the Freeman will usually be by Caesarean section and therefore extensive planning will be required, including theatre provision, neonatal input, maternal after-care and post-natal management strategies.

Lesions which are likely to be associated with significant haemodynamic compromise within the first hour of life, leading to urgent intervention, cardiac surgery or ECMO support, including:

- Hypoplastic left heart syndrome with severely restrictive atrial septum
- Transposition of the great arteries with intact/restrictive atrial septum
- Severe cardiomyopathy (if ECMO likely to be required)

Deliver in Royal Victoria Infirmary:

Cases which are anticipated to require paediatric cardiology input within hours of delivery.

May include:

- Transposition of the great arteries (all forms)
- Duct dependent congenital heart disease
- Functionally univentricular hearts
- Complex isomeric hearts
- High suspicion of coarctation of the aorta
- Common arterial trunk
- Severe Ebstein's anomaly.
- Tetralogy with absent pulmonary valve
- Obstructed anomalous pulmonary venous connection
- Complete heart block
- Uncontrolled fetal tachycardia
- Severe atrioventricular or semilunar regurgitation leading to cardiomegaly (including Hydrops)
- Any critical CHD with spontaneous onset of labour leading to prematurity if transfer is possible and advisable.

Cases delivered in the Royal Victoria Infirmary should have a pre-planned strategy regarding whether the baby will remain in the neonatal intensive care unit or be transferred after delivery to the Freeman Hospital.

Deliver at RVI, James Cook, Middlesbrough, or Sunderland Royal Hospital - whichever is nearest:

Haemodynamically significant lesions, which are unlikely to require urgent paediatric cardiology input, but which should either be seen by a paediatrician with an expertise in cardiology (PEC) or referred for outpatient FRH paediatric cardiology assessment on the day of discharge home.

- Tetralogy of Fallot (with forward flow in the duct)
- Medium/low suspicion of coarctation of the aorta
- Fetal arrhythmia, which has been well controlled with transplacental therapy
- Mild/ moderate aortic valve stenosis
- Double aortic arch

Deliver in booking hospital:

Lesions which are unlikely to require investigation or treatment in the neonatal period – suitable for non-urgent paediatric cardiology or PEC outpatient clinic referral.

Includes:

- Isolated ventricular septal defect
- Partial or Complete AVSD each with balanced ventricles
- Mild/ moderate pulmonary valve stenosis (isolated)
- Mild atrioventricular valve regurgitation
- Right sided aortic arch
- Isolated Left sided SVC
- Bilateral SVCs (isolated)

Post-natal management strategy:

All antenatally diagnosed congenital heart disease should have a documented plan covering presentation to the local unit in preterm labour.

For cases delivering at the Royal Victoria Infirmary:

Prior to induction of labour or at the time of presentation in labour, the maternity team should inform the neonatal team and the consultant paediatric cardiologist on call that the baby is expected. In cases where the fetal cardiology team recommend routine commencement of a prostin infusion, this should be started at a dose of 5 nanograms/kg/minute.

For cases delivering elsewhere:

Local strategies should be in place for the neonatal/paediatric team to be informed once labour takes place/delivery is planned. If there are unexpected neonatal concerns or if for any reason paediatric cardiology advice is required, the neonatal/paediatric team in the delivery hospital should contact the consultant paediatric cardiologist on call, via Freeman Hospital switchboard.

In those units with a local paediatrician with expertise in paediatric cardiology (JCUH, SRH) there is the further option of arranging local review providing this would not result in a significant delay.

At the point of discharge from any hospital, the neonate with confirmed or suspected congenital heart disease should have appropriate review arrangements in place prior to hospital discharge. Where a routine paediatric cardiology outpatient appointment has been agreed, the paediatric cardiology registrar on call should be made aware that a referral is being sent and the on call paediatric cardiologist should be been informed.