

Single Ventricle Pathway

Initial Encounter

- Level II Ultrasound
 - Growth documentation
 - Placental size and appearance
- Genetics
 - Karyotype for 22q- deletion testing
 - If multiple anomalies are present, order limited ARRAY testing.
 - NO ARRAY for isolated CHD
- Fetal Echo: Identify atrial septal restriction
- Parents/mother given information sheet, including info about fetal intervention
- Introduce Perinatal/Neonatal Navigators
- Tour Birth Center/Care Planning



Perinatal Care Navigators

- Consults to be scheduled:
 - 1) Cardiologist 2) Neonatology 3) Cardiac Surgery 4) Genetic Counselor
- Tours of NICU and CVCC
- Mother/parents to be given opportunity to speak with families of other single ventricle patients.
 - Schedule by phoning (612) 813-8800.



Subsequent Visits

*** Patients with restricted atrial septum or intact atrial septum should be explicitly identified as they are high-risk and have a different delivery pathway.**

*** Restricted or intact atrial septum patients to be flagged in NNP Perinatal Book.**

- Start Antenatal testing at about 30-32 weeks
 - Monitor fetal growth at least Q monthly
 - Monitor amniotic fluid volumes
 - Repeat fetal ECHO 6-8 weeks after initial diagnosis
 - Re-evaluate and repeat ECHO 4-6 weeks after 2nd ECHO
 - Fax updated information to designated NICU and NICU Office. NICU fax #: MSP (612) 813-6949, St. Paul (651) 220-7085. NICU office fax #: MSP (612) 813-5910, St. Paul (651) 220-7777
- Mother/parents offered repeat consultation (same cardiologist, surgeon, neonatologist)
- Transfer prenatal care to MPP by 32 weeks
- Mother to relocate to Twin Cities by 36 weeks
- If a c-section has been scheduled, notify the Cardiology Office and NICU





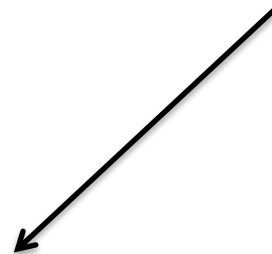
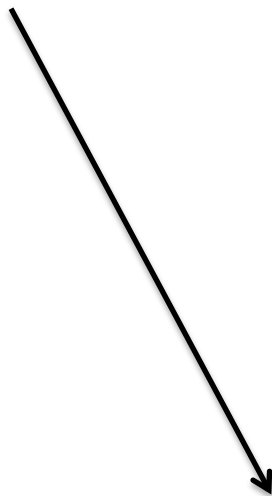
Resuscitation Room

- Delivery at 40-41weeks (Ideal)
- Notify cardiologist and ECHO technologist when NICU team leaves for DR
- Keep SpO2 70-90% (Do not over-oxygenate)
- Intubation and PPV only for SpO2 <65, respiratory distress, or shock
- Start PGE1 @ 0.025mcg/kg/min
- Place UAC and UVC

Resuscitation Room for

Restricted or Intact Atrial Septum

- Delivery by scheduled cesarean section at 38-39 weeks (ideal) for patients with restricted or intact atrial septum.
- Parents can refuse elective c/s after informed consent of potential for poor outcomes related to possible delay in treatment is obtained.
- Notify cardiologist and ECHO technologist when NICU team leaves for DR
- Planned intubation and PPV
- Keep SpO2 70-90% (Do not over-oxygenate)
- Start PGE1 @ 0.025 mcg/kg/min
- Place UAC and UVC
- Plan for transfer to cath lab or OR from DR



NICU Care

- Avoid mechanical ventilation for patients without restricted or intact atrial septum, if possible
- SpO2 70-90%
- PaO2 >30mmHg
- ABG at least 4x/day
- Keep pH > 7.35
- Lactate at least 3x/day
- Continue PGE1 infusion
- Peripheral arterial line if no UAC
- PICC line if no UVC
- NPO until surgery
- TPN
- Daily Hct/Hgb
- Keep Hct > 43%/Hgb > 14.5
- TRANSFER TO CVCC by 3 PM day preceding surgery
- CVCC doctor to call Neonatologist for handoff