MINNESOTA NEONATAL PHYSICIANS P.A.
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Revised 2/2016

Congenital Diaphragmatic Hernia Pathway

Prenatal Diagnosis – VISIT #1

1. Level II ultrasound to include
   a. Cardiac ultrasound (by cardiologists)
   b. LHR at 23-26 weeks
2. Recommend Karyotype and microarray testing
3. Genetic counselor referral
4. Refer to MPLS Fetal Care Coordinators
5. Recommend all CDH patients deliver in an ECMO center

Key Contact Information

Minneapolis
Perinatal Clinic: 612-863-4502
Fetal Care Coordinators: 612-863-9924
Schedule Neonatal Consult: 612-813-6288
Schedule Pediatric Surgery Consult: 612-813-8000

Prenatal Consultations

1. Family receives printed CDH information sheet
2. Genetic consultation
3. Discuss prognostic factors
   a. Gestational age
   b. Chromosomes/anomalies/syndromes
   c. Size of lungs
   d. Right sided more likely to need ECMO, similar survival
4. Neonatologist consultation
5. Discussion of pathophysiology and usual treatments including ECMO and given parent manual
6. Pediatric Surgery consultation
7. Opportunity to speak with other CDH families

Eval for Referral Criteria at 23-26 wk

- Nl. karyotype/no other anomalies
- Liver in thorax
- LHR < 1.0
- Parental interest
- MRI if meets all above criteria

Yes to all

Refer for fetal surgery 23-26 weeks

Subsequent Perinatal Visits

1. Prenatal Care transfer to MPP at 28-32 weeks
2. Monitor monthly for fetal growth
3. Weekly BPP at ≥ 32 weeks
4. Antenatal steroids at 38 weeks, empirically or threatened preterm delivery
5. Convey information to NICU/neonatologists
6. Mother moves to Twin Cities by 36 weeks
7. Goal: Delivery after 39 $^{0/7}$, induce after 40 wks

Follow-up Information

1. Fax updated info to NICU attn NNPs
   a. 612-813-6949 (Fax) MPLS

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Delivery/Resuscitation

1. Vaginal unless other indication
2. Immediate intubation
3. Ventilate with ≤ 30 cm H_2O (PIP 25-30/Peep 5-6). Start with 40% oxygen and increase as needed. Goal is preductal SpO_2 ≥ 85% by 15 minutes
4. Prefer Neo-Puff or ventilator to regulate pressure
5. Start iNO @ 20 ppm, if needing >60% oxygen
6. Place UAC and UVC; UAC – ½ Normal Acetate
7. Blood culture, ABG and lactate
8. NO surfactant unless immature L/S or < 34 weeks *and* no betamethasone
9. NO routine muscle relaxants
10. O/G to suction – 10 Fr Repogle
11. Morphine +/- benzodiazepine for agitation/pain, only as indicated

Special Items to Take to Delivery Room

1. iNO
2. Morphine
3. Muscle relaxant
4. Transilluminator
5. iSTAT machine
6. Repogle OG - 10 Fr

NICU Initial Management

1. Ventilation goals – **Emphasize Gentle Ventilation**
   a. SpO_2 target 90-98% pre and post-ductal
   b. Wean FiO_2 slowly and only if SaO_2 > 90% post-ductal
   c. Maintain pCO_2 < 65 and pH > 7.25
   d. Keep CV PIP ≤ 30 cm H_2O or JET PIP ≤ 45, or HFOV AMP ≤ 45 and MAP ≤ 15 cm H_2O
   e. Use HFJV if PIP > 25 or need more CO_2 removal. Initial HFJV setting 38/8 rate 420
2. iNO (20 ppm) if FiO_2 > .60 and/or evidence of PPHN
3. Cardiac output – monitor lactate. Pressors as needed to support BP and reduce R to L shunting
   a. Dopamine (0-20 mcg/kg/min)
   b. Dobutamine (0-20 mcg/kg/min)
   c. Hydrocortisone (1 mg/kg Q8H) if inotropes needed
   d. Epinephrine 0.01-0.5 mcg/kg/min
   e. Vasopressin for resistant hypotension 0.0001-0.001 units/kg/min (causes marked sodium loss and dieresis)
   f. NO milrinone in acute phase (may be added once BP stable)
4. Central venous catheter and arterial line. If possibility of ECMO, need large bore venous access – UVC or femoral CVC inserted by PICC or CVCC MD, as can't give blood products rapidly via PICC.
5. Sedation/analgesia episodic →drip if needed
   a. Morphine – 0.05 mg/kg q 3-4 hr prn
   b. Midazolam or lorazepam prn
6. Muscle relaxants, only if needed for bowel decompression or severe respiratory failure
7. Position supine or on side of hernia, not on contralateral side
8. Antibiotics (Amp and Gent) for 48 hr pending sepsis evaluation
9. O/G tube to LIS
10. Type and screen and State Newborn Metabolic screen.
11. CGH if not done prenatally and baby: SGA, LGA, FLK, other anomalies, prior fetal loss or child with birth defect, or likely to need ECMO or die
12. Obtain echocardiogram
13. **See ECMO criteria –** separate criteria for CDH

NICU Initial Monitoring

1. Pre and Post Ductal SpO_2 monitors (90-98%)
2. Monitor lactate if on inotropes (should be <20mg/dl)
3. Mean arterial pressure should be > 40 mm Hg, or appropriate for gestational age
4. pCO_2 < 65 and pH ≥ 7.25
5. ABGs 5-6/day and record post-ductal OI in I-view for all ABGs pre repair. If on HFJV be sure to use "Jet MAP".

Criteria for Transfer to ECMO Center (any of the following)

If born at Level I or II center, direct transfer to an ECMO center

1. Oxygenation Index (OI) ≥ 15
2. MAP > 12 cm H_2O
3. Amplitude > 30 cm H_2O or PIP ≥ 25 cm H_2O
4. Plasma lactate > 20 mg/dl and not improving
5. Hypotension (mean art. Pressure < 40 mm Hg) on moderate dose inotrope (dopamine > 10)
6. Acidosis (pH < 7.25 or base deficit > 5) despite dopamine > 10 mcg/kg/min
7. Echo evidence of poor RV function or near

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Criteria for Diaphragmatic Hernia Repair

1. Clearing lung fields
2. Weight to within 120% of presumed dry weight
3. On conventional ventilation
4. $V_T \geq 4$ ml/kg at PIP < 30 cm H_2O and PEEP 4-6 cm H_2O
5. Rt. Vent. Press. ≤ 2/3 systemic, $FiO_2 < 0.40$ and stable BP on dopamine ≤ 10 mcg/kg/min
6. Pre-op antibiotic prophylaxis with oncef
7. Make sure blood available for surgery (1 PRBC units)
8. See ECMO protocols for other items if on ECMO.

Immediate Post-Operative Management

1. No routine chest tube, unless on ECMO
2. Chest tube, if present, to 10 cm H_2O; if no bleeding, water seal
3. Abdominal pressure measurement if requiring PIP > 25 cm H_2O, or urine output < 1.5 ml/kg/hr
4. iNO if $FiO_2 > 0.60$ and/or evidence of PPHN
5. Analgesia
6. See ECMO Pathway for CDH patients on ECMO

Chylothorax

1. Follow Anti-thrombin III levels. Keep 60% if CVC present
2. Follow Albumin levels. Keep > 2.0.
3. Replace output with albumin for volumes < 25 ml/kg/day. FFP for volumes > 25 ml/kg/day. Replace ~ 50% of output.
4. See Chylothorax Management Guideline

Intra-abdominal Pressure (IAP) Management

1. Monitor IAP at least hourly (Should be < 15 mm Hg)
2. Notify MD if IAP > 15 mm Hg and consider
   a. Remove constrictive dressings
   b. Elevate head of bed to 30 degrees
   c. Neuromuscular blockade
   d. Diuresis/hemofiltration
   e. Vasoactive meds to keep perfusion pressure adequate
3. For IAP > 20 mm Hg, which is the definition of intra-abdominal compartment syndrome and/or urine output < 1 ml/kg/hr, or worsening acidosis, strongly consider surgical intervention

Long-Term Management

1. Persistent pulmonary hypertension
   a. Continue iNO weaning **slowly,** monitoring for rebound with echocardiogram after all major changes (i.e. discontinuing iNO).
   b. Consider transition to oral sildenafil when on feeds if unable to wean iNO to off. Start at 0.5 mg/kg Q6H and advance to 2 mg/kg Q6H
   c. SpO_2 targets 92-98%
   d. Do not wean below 30% oxygen until off iNO and sildenafil and echo shows minimal pulmonary hypertension
2. Other
   a. Regular feedings (MBM or formula) unless documented chylothorax
   b. Start with continuous feedings and switch to Q3H bolus feedings
   c. Position HOB elevated 30 degrees. If symptomatic GER start proton pump inhibitor
   d. Wean morphine/other narcotics by 5-10% per day
   e. Repeat State metabolic screen when off steroids for 10 days if indicated.
   f. Consider OT consult and GJ tube for poor oral feeding.

Plan for Post-Discharge Care

1. Pulmonology follow-up for oxygen dependent CLD, or pulmonary hypertension
2. Cardiac ECHO prior to discharge if any concern of ongoing pulmonary hypertension
3. Cardiology consult and outpatient follow-up if RVSP > 25 mm Hg or other evidence of continued pulmonary hypertension
4. MRI prior to discharge if received ECMO
5. Hearing screen every 6 months until 3 years and then annually until 5 years of age
6. Careful growth monitoring in first year of life
7. Consider Danny sling with HOB elevation and proton pump inhibitors for symptomatic GE reflux
8. Follow-up with pediatric surgeon in 2-3 months
9. NICU Follow-up Clinic evaluation at 9, 18, 36 months
10. Consider Feeding Clinic Referral if ongoing problems or growth failure
11. Recommend screening for scoliosis by primary care until adult weight and height
12. Influenza vaccine for family and for patient (after 6 months old).
13. RSV prophylaxis for all CDH patients on oxygen after discharge, in first winter