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Management of Congenital Diaphragmatic Hernia (CDH) –

Neonatal – Inpatient - Guideline Summary

Target Population: Neonatal patients with congenital diaphragmatic hernia

Link to Full Guideline: Management of Congenital Diaphragmatic Hernia (CDH) - Neonatal - Inpatient

## Analgesia and Sedation

- Analgesia and sedation should be provided and monitored with validated scales (SBS or N-PASS)
  - Initiate dexmedetomidine first line (starting dose 0.2-0.3 mcg/kg/hr)
  - Use morphine as needed for additional analgesia (starting dose 0.01-0.02 mg/kg/hr), with goal of using the minimal necessary dose
  - o Midazolam can be considered for additional sedation if clinically necessary (after optimizing other agents)
  - Paralysis and deep sedation should generally be avoided; may consider for patients requiring maximal respiratory support • Deep sedation using midazolam and morphine is required when administering paralysis

Pulmonary Hypertension Management Considerations	Hemodynamic Management Considerations
<ul> <li>Acute PHTN treatment is recommended if ≥1 of the following:         <ul> <li>Pre-ductal SpO2 &lt; 90% despite ventilatory optimization</li> <li>Oxygenation index (OI) &gt; 25</li> <li>Post-ductal sats &lt; 70% and/or evidence of end organ dysfunction</li> <li>Echo with systemic or near-systemic right-sided pressures and depressed RV function <u>or</u> if evidence of supra-systemic to near-systemic right-sided pressures with borderline oxygenation.</li> </ul> </li> <li>A trial of iNO should be initiated as the 1st line treatment for confirmed PHTN; have caution if significant LV dysfunction</li> <li>Prostaglandins may be useful in the specific circumstances         <ul> <li>See <u>full guideline</u> for detailed considerations</li> </ul> </li> <li>Sildenafil may be considered in refractory PHTN (typically late or subacute phase) or if longer-term treatment is necessary</li> </ul>	<ul> <li>For <u>hypovolemia</u>, use isotonic crystalloids judiciously</li> <li>For <u>hypotension</u>, consider the following to increase SVR and BP in the setting of high PVR and good RV function:         <ul> <li>Norepinephrine (dosing range 0.05-1.0 mcg/kg/min)</li> <li>Epinephrine (dosing range 0.01-0.1 mcg/kg/min)</li> <li>Vasopressin (dosing range 0.17-10 milliUnits/kg/min)</li> <li>Dopamine may worsen PHTN and should be considered with caution (dosing range 5-25 mcg/kg/min)</li> </ul> </li> <li>For <u>RV dysfunction associated with PHTN</u>, milrinone is the preferred 1st line agent</li> <li>For <u>ventricular dysfunction</u>, dobutamine (dosing range 5-20 mcg/kg/min) is an additional therapeutic option</li> <li>Consider hydrocortisone (1 mg/kg Q8 hrs) to address adrenal insufficiency and hypotension</li> </ul>

## **ECMO Considerations**

• There are no specific criteria for ECMO, the indications and contraindications below may be used to determine eligibility.

- If baby is meeting criteria for ECMO, early discussion with NICU, PICU and pediatric surgery attendings should occur.
- Echo and Cranial US are necessary for evaluation, but their completion should not delay transfer

Criteria for ECMO Consideration	Absolute Contraindications	Relative Contraindications
<ul> <li>Prenatal indicators for ^likelihood of ECMO need:         <ul> <li>O/E TFLV &lt;25% or PPLV &lt;15%</li> <li>O/E LHR &lt; 25%</li> <li>liver up</li> </ul> </li> <li>Postnatal indicators prompting ECMO consideration when sustained and refractory to treatment:         <ul> <li>Preductal SaO<sub>2</sub> &lt; 85%</li> <li>PaCO2 &gt;70 and/or pH &lt;7.20</li> <li>OI &gt; 40 [OI = (FiO<sub>2</sub> x M<sub>PAW</sub>) / PaO<sub>2</sub>]</li> <li>PIP &gt; 30 or HFOV AMP &gt;45</li> <li>lactate &gt; 4 mmol/L</li> <li>Circulatory failure                 <ul> <li>Severe hypotension</li> <li>LV failure</li> <li>Severe PHTN resulting in RV failure</li> </ul> </li> </ul> </li></ul>	<ul> <li>Gestational age &lt; 32 weeks</li> <li>Birth weight &lt; 1.6 kg</li> <li>Lethal comorbidity (Pentalogy of Cantrell, Frynn's syndrome, Bilateral defects)</li> <li>IVH ≥ grade 3</li> </ul>	<ul> <li>Gestational age 32-34 weeks</li> <li>Birth weight &lt; 2 kg</li> <li>IVH grade 2</li> <li>Cardiac lesion (coarctation)**</li> <li>Failure to achieve SaO<sub>2</sub> &gt; 85% (and sustain for 1 hour) within the first 2-4 hours of life despite maximal ventilatory maneuvers</li> </ul>

## Surgical Repair Considerations

The timing and approach to surgical repair should be individualized. Infants should generally undergo repair following clinical stabilization – consider the following physiologic criteria when evaluating stability: OI < 9, urine output > 1 mL/kg, FiO2 < 0.5, preductal sats 85-95%, normal MAP for gestational age, lactate < 3 mmol/L, and estimated PA pressures < systemic pressure.

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## Ventilator Management and Related Care Measures for Infants with CDH

