

# Congenital Diaphragmatic Hernia: Management Guidelines 5-2006

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## Congenital Diaphragmatic Hernia: Management Guidelines

We may best reach the goal of intact survival for the most infants with congenital diaphragmatic hernia (**CDH**) with a strategy geared toward minimizing lung injury and careful utilization of ECMO. Unfortunately, measures of injury and adequate perfusion, oxygenation, and ventilation at the cellular level are lacking. Arbitrary cutpoints indirectly related to these more relevant facts will be used to guide management.

Patient care will be followed by representatives from pediatric surgery and neonatal medicine. Individual case reviews will follow the care of babies with **CDH**. The guidelines will be adapted to optimize clinical outcome based on our experience and published outcome data from other centers. Recent changes include mention of early use of iNO in reactive pulmonary hypertension.

### Summary of Therapies

Support will be described in terms of safe, caution, and hazard zones. Patients will be described as "ideal" or "non-ideal acceptable." Particular therapies and situations are discussed in more detail in subsequent sections.

#### Safe Zone:

CV PIP < 26; HFOV: MAP < 16; HFJV: MAP < 16

Dopamine infusion up to 20 mcg/kg/min

Epinephrine infusion up to 0.1 mcg /kg/min.

#### Caution Zone:

CV PIP 26 - 30; HFOV: MAP 16 - 22; HFJV MAP 16 - 22

Dopa 20mcg/kg/min + Epinephrine 0.1 - 0.3mcg /kg/min

Alkalinization

iNO

#### Hazard Zone:

CV PIP > 30; HFOV MAP > 22; HFJV MAP > 22

Dopa 20mcg/kg/min + Epinephrine > 0.3 mcg /kg/min

Ideal patients maintain O<sub>2</sub> saturations > 95% with safe zone ventilator and vascular support limits. If this is not achieved, non-ideal acceptable patients may be medically managed with PaCO<sub>2</sub> up to 65 mmHg and postductal PaO<sub>2</sub> as low as 30 mmHg, as

long as pH remains greater than 7.25 without evidence of worsening acidosis/rising serum lactates and preductal O2 saturations are > 85%. If a baby requires caution zone therapies to maintain ideal patient parameters (and our strategy is to limit lung injury) we will assess duration of such support and consider weaning, particularly ventilator support, with a goal of non-ideal acceptable patient parameters. If the baby needs caution zone therapies longer than 24 hours to maintain non-ideal acceptable parameters, neonatology, pediatric surgery, and ECMO service attendings will discuss time limits for current therapies and endpoints leading to ECMO. If hazard zone treatments are required longer than a few hours to maintain the patient in the ideal range, weaning to caution or safe zone treatments to reach acceptable non-ideal patient standards should occur. If this is not possible and hazard zone treatments are required to reach and maintain acceptable non-ideal patient standards, ECMO must be considered.

### Antenatal management

Once a fetus with **CDH** is identified we recommend prenatal assessment of cardiac anatomy with fetal echocardiography. Biophysical profiles should be performed as clinically indicated. Pediatric surgery and neonatal medicine attendings should meet with the families to discuss care and outcome of patients with **CDH**. Pediatric surgery attendings will review options regarding fetal surgery. Obstetric patients and their infants should be enrolled in the **CDH** Study Group registry. We will maintain our own **CDH** database for further analysis of the proposed guidelines.

### Antenatal steroids

Expectant mothers identified with a fetus with **CDH**, and threatened delivery prior to 34 weeks gestation should be offered antenatal steroids. There is no current indication for antenatal steroids in late preterm or term **CDH** infants.

## **Initial Management**

### At delivery

**CDH** infants demand immediate intubation and placement of an adequately sized Replogle tube (ideally at least 10 French). Initial positive pressure ventilation PIP's ideally will not exceed 24, although for initial breaths, and I would stress the word initial, higher inspiratory pressures may be necessary. Pressures between 24 and 30 should be considered aggressive, and for very short-term use.

Early use of ventilators rather than hand bagging patients offers information about tidal volumes and compliance, as well as consistent pressures and rate. The ventilator should be used as early as possible.

Radiographs must be used to verify tube positions (ETT and Replogle, as well as umbilical lines), and distances recorded in the patient record.

**CDH** infants should be placed on a bed scale.

### Surfactant

We do not include early surfactant for term born **CDH** infants in standard practice, but after initial resuscitation, extremely cautious dosing can be considered based on clinical indications.

### Ventilatory Management

Initial goal is tidal ventilation (conventional ventilation, CV) and pre- and post-ductal O<sub>2</sub> sats > 95%. If PIP > 26 and/or MAP > 12 are necessary to achieve PaCO<sub>2</sub> < 65 mmHg, HFV will be used.

HFV: Start HFV if PIP > 26 with CMV is necessary to maintain ideal patient parameters. Initial HFV will be with jet ventilation.

Safe zone: CV PIP < 26; HFOV: MAP < 16; HFJV MAP < 16

Caution zone: CV PIP 26 - 30; HFOV: 16 - 22; HFJV, MAP 16 - 22

Hazard zone: CV PIP > 30; HFOV MAP > 22; HFJV MAP > 22

### Ventilator Weaning

For ideal patients with safe zone settings, weaning may begin after achieving ideal blood gases. FiO<sub>2</sub> should be decreased by no more than 0.03/hour, with the goal of maintaining pre and post ductal O<sub>2</sub> saturations > 95%. Weaning of pressures (MAP on HFOV, PIP and PEEP on conventional and HFJV) and rate may begin after weaning to FiO<sub>2</sub> of 0.40 - 0.50. When evidence (CXR's, volume loops) shows hyperinflation or blood gases reveal hyperventilation, pressure and rate weaning may be accelerated. Ideal patients with safe zone settings need not be weaned to reach acceptable, non-ideal blood gas/O<sub>2</sub> saturations. If the lung appears well inflated, weaning settings to find the lower limit of pressures to maintain ideal parameters is encouraged.

Patients with ideal gases/SaO<sub>2</sub> and hazard zone settings must be weaned.

If while weaning the infant's SaO<sub>2</sub> and blood gases drop from ideal to non-ideal acceptable (preductal sat > 85%, pH > 7.25) and the drop is not secondary to a loss of lung volume, consider not increasing ventilator settings, unless the settings started in the safe zone. If settings are within the safe zone a return to previous support is acceptable. If evidence (CXR, loss of chest rise, decreased exhaled volume) points to loss of lung volume for the drop, ventilator settings may be increased into caution zone, and with extreme situations for a limited time, hazard zone. If the wean was from caution zone to safe zone, and the patient falls from ideal to acceptable, consider staying at the safe zone support level. If pressures in the caution range (CV PIP 26 - 30; HFOV MAP: 16 - 22; HFJV, MAP 16 - 22) are necessary to reinflate and maintain lung volume for longer than 24 hours, ECMO must be considered.

### Sedation

Keep the environment quiet!

In the Unit: bolus dose of 50 mcg /kg morphine followed by a morphine drip at 10 mcg/kg/hour, with intermittent bolus doses of fentanyl or morphine, titrated to needs, of 5mcg /kg fentanyl or 50 mcg /kg morphine. Be prepared for volume and/or inotrope support after bolus doses

In the Unit (alternative): Midazolam drip at 3 mcg/kg/minute, with morphine boluses titrated for anticipated pain, starting with 50 mcg/kg/dose. Be prepared for volume and/or inotrope support after bolus dosing.

Beware of possible hypotension/arrest with combination of opiates and midazolam!

Beware of hypotension, possible hypoadrenalism with opiate infusion (not studied with midazolam)

### Surgery criteria

Surgery in the first few days of life is encouraged for infants requiring safe zone support. A period of 24-48 hours at this range of support is desirable.

### Blood pressure management (with or without PPHN)

Optimize ionized calcium, check cortisol level and consider hydrocortisone stress doses if dopamine up to 10 mcg/kg/min has not achieved or maintained acceptable mean arterial pressure (MAP) or cortisol level is low. Optimize hematocrit 40 - 50% as polycythemia may lead to pulmonary hypertension.

MAP for term infants with **CDH** without significant pulmonary hypertension and not needing significant respiratory support will be acceptable as low as 40, as long as tissues are well perfused, urine output is adequate, and post-ductal pH is > 7.25 with a low serum lactate, and the patient is otherwise ideal and requires safe zone respiratory settings.

When cardiovascular performance is suboptimal, pressors will be added. We will start with 5 mcg /kg/min of dopamine.

This can be increased to 20 mcg /kg/min, where, if MAP continues inadequate, hydrocortisone will be added (stress dose if the initial cortisol level is low, maintenance dose if the level is high). Weaning off hydrocortisone may lead to adrenal insufficiency, with acute rises in potassium, BUN, and creatinine, and oliguria.

Volume pushes are avoided unless there is evidence of decreased intravascular volume.

Post-operative diuresis should be carefully followed. Unless oxygenation is compromised and/or ventilator weaning is limited due to excessive fluid, diuresis should be gradual achieving negative fluid balance of no greater than 100-200 ml/day, while carefully following measures of intravascular volume (weight, electrolyte, BUN and creatinine, vital signs).

If dopamine plus the above does not provide relief, epinephrine will be our second agent, and will be started at 0.05mcg /kg/min.

### Persistent Pulmonary Hypertension (PPHN) management

Blood Pressure:

When trying to exceed pulmonary vascular resistance, we will aim for MAPs up to 60. Refer to the general guidelines for review of safe, caution, and hazard support guidelines.

Therapy used to reach these goals includes calcium, hydrocortisone, dopamine, and epinephrine.

Hematocrit should be optimized to between 40 and 55%.

Dopamine doses will be limited to 20 mcg/kg/min, and will be considered "safe zone" therapy.

For epinephrine doses:

Safe range: <0.1 mcg/kg/min.

Caution zone: 0.1 - 0.3mcg /kg/min.

Hazard zone: > 0.3 mcg /kg/min. Assessment of hazard zone therapy with epinephrine must include assessment of cardiac function.

### iNO

NO should be considered a caution zone therapy. In **CDH** cohort studies, and **CDH** subgroup analysis of the NINOS trial, iNO did not reduce mortality or ECMO risk. However, in individualized cases where reactive pulmonary hypertension may be causing hypoxia and could lead to acidosis and further fixed pulmonary hypertension, iNO may benefit individual patients.

For the "acceptable" patient on safe or caution zone settings, if pulmonary hypertension is evident by cardiac echo/saturations, iNO may be tried. If there is no response after 1 hour, the iNO should be weaned and discontinued (if possible). If pulmonary hypertension persists, and therapy has increased to include epi- and intent to alkalinize, iNO may be tried again.

If the patient has preductal O<sub>2</sub> saturations < 85% after the initial hours of management, and has pulmonary hypertension, iNO may be used to attempt to bring the patient to acceptable levels, or as a bridge to ECMO.

### Alkalinization

Alkalinization should be considered a "caution zone" therapy.

In general, sodium bicarbonate and THAM will be used only to treat metabolic acidosis. If cardiac echo and clinical condition suggest pulmonary hypertension and echo shows good right and left ventricular function, and the patient's O<sub>2</sub> saturations and measures of perfusion are less than ideal, medical (rather than ventilatory) alkalinization may be used to achieve ideal patient parameters. The goal for pH will be up to 7.60. There are no studies of the relative efficacy of alkalinization for these patients vs. use of iNO.

Once alkalinization is achieved, if more than safe zone ventilator settings or pressors are needed along with alkalinization to maintain ideal patient parameters, consider reducing ventilator and pressor support and to obtain adequate patient parameters. This may reduce the risk of iatrogenic morbidity.

### ECMO criteria

Inability to consistently maintain postductal PaO<sub>2</sub> of 30 mm Hg, preductal O<sub>2</sub> saturation of 85% or post-ductal pH < 7.25 suggest failed medical management.

If the combination of iNO or alkalinization + hazard zone pressor or hazard zone ventilator settings are necessary to achieve and maintain acceptable non-ideal blood gases and SaO<sub>2</sub>, this should be considered a bridge to ECMO.

If pressors, vent support or alkali are in the caution range, i.e. epi 0.1- 0.3 mcg/kg/min, CV and HFJV PIP 26 -30 (32 for HFJV), alkalinization requiring pH 7.5 - 7.60, and/or nitric oxide have been used to achieve at best acceptable non-ideal patient parameters, some agreement of time limit for this management should be set by the combined efforts of pediatric surgeons and neonatologists.

Reaching OI's of >40 x 2 in one hour should lead to consideration of ECMO. (MAP 20 on HFOV, FiO<sub>2</sub> 1.0, and PaO<sub>2</sub> of 50 gets you an OI of 40).

In addition to O<sub>2</sub> saturations and a pH guideline of 7.25, serum lactates on the rise

should lead to close scrutiny of present and the immediate hours of past management.

### Patient monitoring

Pre- and post-ductal O<sub>2</sub> saturation monitors.

Transcutaneous CO<sub>2</sub> monitors are especially necessary for those on HFJV or HFOV to avoid hyperventilation.

Pre- and post- ductal arterial access (ideally).

For non-ideal patients or ideal patients on caution or hazard level support:

Frequent (at least q6) measures of serum lactate will be one of the few available measures of tissue perfusion.

Frequent (q blood gas) ionized calcium

Q 4 blood gas (minimum)

### Echocardiography

For the ideal patient, only one may be required to assess anatomy. Include a modified Nakata index (hilar PA diameter/surface area) a modified McGoon index (hilar PA diameter/descending aorta diameter) as per Suda et al, Pediatrics 2000;105:1106 - 1109. (Discussed with Dr. Andersen, Pediatric cardiology)

For any patient to receive iNO, a cardiac echocardiogram during iNO administration will be helpful to assess source of pulmonary hypertension (and help guide subsequent therapy).