ANTENATAL MANAGEMENT

Due to the increased availability of prenatal ultrasound, 60-80% of CDH cases are now diagnosed antenatally. Referral to the Maternal Fetal Medicine group is desired whenever possible, to allow a full evaluation of the fetus. In addition, coordination of the multiple consultants required for care is best done prior to delivery, as this allows development of a comprehensive plan for delivery as well as providing the family with information at an optimal time.

ANTENATAL EVALUATION:

1. Ultrasound

   The ultrasound is used to:
   - Confirm the diagnosis, and determine if L or R sided
   - Identify associated anomalies
   - Assess for increased amniotic fluid or the presence of hydrops
   - Assess the position of liver (in abdomen or herniated into the thorax)
   - Assess the position of stomach (in abdomen or herniated into the thorax)

   The use of lung volumes or the lung to head ratio (LHR) is still being developed and should not be used for prognostication at this time. The use of 3D Ultrasound is still a novel technology which may prove useful. At the current time it should not be considered standard of care. The position of the stomach is useful for diagnosis, but should be used with caution for prognostication. It is recommended to obtain repeated scans until delivery.

2. Fetal Echocardiography

   Fetal echocardiography is helpful in assessing:
   - The presence/absence of structural anomalies
   - The size of RV and LV
   - The cardioventricular index: LV / RV
   - The cardiovascular index: Aorta / Pulmonary artery
   - Fetal PA measurements especially Left Pulmonary artery measurement

   It is recommended to obtain an initial ECHO and then at least one follow-up ECHO in the third trimester to assess the above indices.

3. Ultrafast Fetal MRI

   Fetal MRI evaluation for CDH is a new technology. As knowledge is developed, there will be a better understanding of how to use the data obtained for prognostication. Currently, it is not clear how to interpret discrepancies between ultrasound and fetal MRI. The estimation of the volume of liver herniated into the thorax and of lung volumes is not well established.
However, MRI can be used to better determine associated anomalies and to better differentiate between CDH and other lung pathology, such as CCAM.

4. **Karyotyping**

Amniocentesis for karyotyping should be offered as a high percentage of fetuses with CDH have associated chromosomal defects.

5. **Discussion by the Multidisciplinary Team**

Early involvement by all members of the multidisciplinary team is ideal, to allow for coordination of investigations, counseling and planning of the delivery and immediate postnatal care.

Members of the team should include:
- Obstetrician
- Maternal Fetal Medicine Specialist
- Geneticist
- Paediatric Surgeon
- Pediatric Cardiologist
- Neonatologist
- Social worker

Based on anomalies found during the assessment of the fetus, other members may need to be involved.

6. **Fetal Surgery**

Fetal surgery is still regarded as experimental. Toronto is the only centre in Canada where this is performed. This option should be discussed with families as needed.

7. **ECMO**

ECMO for Neonatal CDH patients is currently not available here in Calgary. The Edmonton ECMO program does not recommend the use of ECMO in these patients. Thus, there is no advantage for the baby to be delivered in Edmonton, unless there is a serious co-existent cardiac lesion. ECMO is infrequently used for CDH patients in other centers in Canada.

8. **Discussion with the family**

Discussions should occur with the family as soon as a diagnosis can be confidently made, and should include:
- Information regarding the prognosis.
- Therapeutic options, including termination, continuation of the pregnancy, the availability of fetal therapy.
- The need to deliver at a tertiary centre (FMC)
- Explanation regarding the lack of advantage of early delivery or elective C/section
- Anticipated delivery of a term infant with CDH, therefore no indication for antenatal steroids
- A joint consultation with members of the multidisciplinary team and the family may be optimal.
RESUSCITATION

1. Anticipation and preparation are vital.

2. Delivery should occur in a tertiary centre.

3. A neonatologist ought to be present at the delivery, if possible.

4. Intubate immediately – ensure that the ETT is correctly positioned ie, that the ETT tip is not in the R main stem bronchus which can lead to over distension of the R lung and a pneumothorax. (The tip-to-lip measurement for ETT placement is 6 + weight in kg as the cm mark at the lip – approx. 9 cm at the lip for a full term baby)

5. Avoid bag and mask ventilation which can cause over-distension of the stomach / bowel

6. Use 50% oxygen at the beginning.

7. Connect the baby to mechanical ventilation. This results in better control of ventilating pressures compared to “hand-bagging”. Neopuff may be used in the absence of a ventilator.

8. Apply an oxygen saturation probe in a preductal location (e.g., right wrist) and use a pulse oximeter for monitoring.

9. Use principles of Gentle Ventilation from the beginning aiming to keep PIP < 25 cm H2O.
   - Initial settings: PIP= 20 -22 cms           Insp Time = 0.35
     PEEP= 3 – 5 cms                       Exp Time = 0.65
     MAP= < 12 cms                        RATE= 20 – 40 / min (Max 60/min)
   - Avoid vigorous chest rise
   - Resuscitation of a baby with CDH should aim at getting a stable heart rate and gentle chest movement recognizing that initially the baby may remain cyanosed.
   - The key principles of successful delivery room resuscitation and stabilization are based on the avoidance of high airway pressures and the establishment of a satisfactory preductal arterial saturation (>75-85%).

10. Decompress the stomach with an O/G or N/G tube (preferably a Replogle tube) and transfer to NICU.

11. Establish arterial access and central venous access in the NICU. Draw blood gas immediately.

12. The use of neuromuscular blockade is discouraged.
Equipment Needed at Delivery

- Bagging unit
- Endotracheal tubes, laryngoscope.blades
- Suction catheters, wall suction
- OG/NG tubes, Replogle tube
- Syringes
- Pulse oximeter, pulse oximetry probes, cable (max sensitivity setting, 2 second averaging times)
- Peripheral iv’s, UAC, UVC
- Sterile gloves, chlorhexidine
- Procedure trays for central line insertion
- Chest tubes
- Mechanical ventilator and necessary tubing
- Neopuff
- Expired CO2 detector
- Medications: fentanyl, atropine, succinylcholine, normal saline, 4.2% sodium bicarbonate, epinephrine

Personnel Needed at Delivery

- Neonatologist
- Respiratory therapist
- Nurses (2 senior)
- Senior bedside clinician (Neonatal Fellow/NNP).
- Optimum number of personnel is 5. An additional person in a learner role may be permitted at the discretion of the Neonatologist/senior clinician.
VENTILATORY MANAGEMENT

The two key factors in determining the outcome of CDH are the degree of pulmonary hypoplasia and increased pulmonary vascular resistance (PVR). Surgical reduction of the hernia and repair of the defect, although an essential component of the overall management has little influence on the outcome and, if done too soon, may jeopardize the infant’s survival. The principles therefore of preoperative stabilization are to optimize ventilation, without causing further pulmonary barotrauma, and to reduce PVR.

The most important principle in the management of CDH is “gentle ventilation and permissive hypercapnia” along with preservation of spontaneous respiration, especially avoiding neuromuscular blockade, if possible.

1. **Mode of Ventilation**

   The main aim is to keep the PIP < 25 cmH2O with the objective of maintaining a preductal O2 saturations of > 85% while tolerating hypercarbia.

   (a) **SIMV mode:** Ventilation should begin in the SIMV mode with the following settings.

   ```
   Initial settings:
   PIP = 20 - 22
   PEEP = 4 – 5
   MAP = 8 – 12
   Rate = 20 – 40/min
   Ti = 0.30-0.35 sec
   Fi02 = 50 % - 100%
   ```

   Considering the small lung volumes in these infants, aim for tidal volume = 3 – 4 mls/kg.

   If oxygenation is still a concern (i.e. preductal oxygen saturation are < 85%), PIP can be increase to a maximum of 25.

   (b) **High Frequency Ventilation:** HFOV should be consider when the PIP on conventional ventilation is > 25 cmH2O and preductal O2 saturation is < 85% or PaCO2 > 65mmHg.

   ```
   Initial settings on HFOV:
   MAP = 12 cmH2O (or 1 cm above the MAP on conventional ventilation)
   Hz = 10
   Amplitude = 30 – 35
   I:E Ratio = 33%
   FiO2: 50 % - 100%
   ```

As the lungs are hypoplastic, lung recruitment strategies should be avoided as this increases barotrauma / volutrauma to the lung.

Constant supervision at the bedside is critical when making ventilatory changes. It may take the baby some time to adjust to these changes – as much as possible we should allow adequate time for this adjustment.
Because infants with CDH have very delayed cardio-pulmonary transition, they may take several hours to attain optimal oxygenation. Use preductal oxygenation to guide oxygen requirements and to adjust the ventilator settings. We should try to resist escalating respiratory support during this period if at all possible. In the first few hours of life, it is acceptable to allow the following preductal saturation rather than escalating therapy.

- 0-2 hours- preductal oxygen saturation > 70%
- 2-4 hours- preductal oxygen saturation 75 – 85%
- > 4 hours- preductal oxygen saturation > 85%

Subsequently, the “ideal” preductal O₂ saturations should be 90 - 95%. However, preductal saturation of ≥ 85% is acceptable, as long as the baby has stable blood pressure with normal lactate and no acidosis, indicating normal postductal perfusion.

1.2 Target Blood Gases:

- PH = 7.25 - 7.35
- PaCO₂ = 45 – 65 mm Hg
- No metabolic acidosis (e.g. Base Deficit < 10 and improving)
- Normal lactate levels (< 2)

If the arterial blood gas is from the preductal (i.e. right radial arterial line), consider aiming PaO₂ between 60 – 80 mm Hg. We should avoid using umbilical arterial (postductal) PaO₂ to guide oxygen therapy as long as blood gases shows normal lactate and no acidosis.

1.3 Weaning:

Maintain preductal O₂ saturations 90%-95%. Wean FiO₂ slowly as tolerated. Ensure that the baby is comfortable and other organ function is adequate. Begin to reduce pressures when FiO₂ < 70% with acceptable blood gases:

- PH > 7.25
- PaCO₂ < 60 mmHg
- FiO₂ < 70%

1.4 Surfactant:

Use of exogenous surfactant in infants with CDH has not been shown to have a beneficial effect on survival, need for ECMO or incidence of CLD. Surfactant therapy was associated with lower survival rate and surfactant is not currently recommended in the management of CDH. However, if the baby is preterm and is thought to have some degree of surfactant deficiency, the use of surfactant may be considered. However, extreme caution should be exercised when surfactant is being administered as many CDH babies have a sudden and severe decompensation with surfactant administration.
1.5 Endotracheal Suctioning:

Minimal handing is necessary for these infants and there should be no routine endotracheal suctioning. Suctioning should be done as clinically indicated. Closed in-line suction should be used for clearance of secretions as in all mechanically ventilated infants. (Refer to Regional Child Health Procedure and Policy for Endotracheal Suction 2-S-4)

CARDIOVASCULAR MANAGEMENT

Pulmonary Hypertension

Clinically significant pulmonary hypertension (PH) is an almost universal finding in congenital diaphragmatic hernia (CDH). The physiological bases for PH in CDH are many and include decreased number of pulmonary arterial structures, over-muscularization of terminal vessels of the hypoplastic lung and failure of postnatal modeling. Based on both human and animal studies, pulmonary arterioles in CDH are unresponsive to nitro vasodilators and over express endothelin receptors. Hence, pulmonary hypertension in CDH may have both functional and fixed components which, at least initially, may be unresponsive to therapy. It is, therefore, not surprising that the management of pulmonary hypertension in CDH remains a challenge. While management of resuscitation at birth and ventilation in CDH has become more or less uniform across different centers, management of pulmonary hypertension remains controversial.

Although a number of therapeutic agents are available for the treatment of PH in neonates, information on their use in CDH is based mainly on observational studies rather than randomized trials. Indeed, there is paucity of safety/efficacy data for most therapies in CDH. However, despite lack of evidence, survival and outcomes in CDH may be improved with a standardized approach to care.

Diagnosis of PH and echocardiograms (ECHO)

1. Although PH can be diagnosed clinically e.g. > 5% gradient in the pre and post ductal oxygen saturations, ECHO plays an important and helpful role in the management of PH and is useful in not only confirming the diagnosis and severity of PH but also in ruling out any associated cardiac anomalies. However, it cannot be over emphasized that the use of ECHO in the management of PH should be in conjunction with clinical signs and symptoms. All infants with CDH should have an ECHO after birth. If the mother is scheduled for a C-section or is in labour, cardiology should be notified about the impending delivery. Following delivery cardiology should be consulted. Depending on the clinical status of the infant, the timing of the first ECHO and consult can vary but should happen within 24 h after birth. Indications for an earlier ECHO may include concern for a congenital heart lesion, refractory pre-ductal hypoxemia, refractory hypotension, and a > than 5% gradient in the pre and post ductal oxygen saturations, amongst others.

2. During the first few days after birth an ECHO may be indicated daily and should be done based on clinical status and after discussion with the pediatric cardiologist. However, after the first few days the frequency of ECHOs is likely to decrease and should again be based on clinical indications.

3. Ideally, an ECHO should be done prior to starting any treatment e.g. iNO, prostins. However, one should not wait for an ECHO before starting these therapeutic interventions.

4. The requirement for a repeat ECHO prior to transfer to ACH should be discussed with cardiology.
**Inhaled Nitric Oxide (iNO)**

1. Indications for starting inhaled nitric oxide (iNO) are highly variable across studies. Criteria for starting iNO may include suprasystemic pulmonary pressures, RV pressures 2/3 or 3/4 of systemic blood pressures, refractory preductal hypoxemia and a pre and postductal oxygen gradient of > 5% and an oxygen index of > 25. It is not essential to have an ECHO before starting iNO, although it would be preferable.

2. The starting dose of iNO should be 20 ppm.

3. If there is no improvement in oxygen saturations after starting iNO, it should not be stopped abruptly. Even without an increase in oxygen saturations, iNO can improve RV function, which can be confirmed by an ECHO.

4. If there is a response, as suggested by increasing oxygen saturations and/or decreasing FiO2 requirements, weaning of iNO can be considered at the following parameters: FiO2 < 0.6 with a preductal PaO2 > 60 mmHg and MAP of ≤ 12 cmH2O. Whether there is a response or not, iNO should be weaned gradually. During the weaning process, especially if there has been a response, ECHO should be considered to assess the pulmonary pressures. One can wean relatively quickly initially but more slowly when the iNO is < 5 ppm. Avoid discontinuing iNO at night. Be ready for rebound pulmonary hypertension when the infant comes off iNO, in which situation, iNO may have to be restarted.

**Prostaglandins**

In some centers, prostaglandins are being used as first line agents to treat PH in CDH. Prostins can be used in the following three scenarios:

1. In infants who on antenatal screening have been categorized to the moderate or high risk group e.g. liver in the thorax.

2. Infants who are stable may start to deteriorate when the ductus starts to close. This will be manifest by preductal desaturations and evidence of right heart failure.

3. In situations where left ventricular function is so poor that systemic blood flow may be dependent on the RV output across the ductus.

If starting shortly after birth, the starting dose of prostin should be 0.01 µg/kg/min IV. Higher doses may be needed if reopening the duct is required. While it is preferable to obtain an ECHO before starting prostins, it is not essential. Be ready to manage hypotension when prostins are started.

Weaning of prostins should be done after consulting cardiology.

**Sildenafil**

**Sildenafil should be used only after consultation with cardiology.**

There are three clinical scenarios where sildenafil may be useful:

1. In infants unresponsive to iNO, as sildenafil has a different mechanism of action as compared to iNO.

2. In rebound pulmonary hypertension following weaning of iNO.
3. In infants who are extubated or close to extubation but continue to have evidence of pulmonary hypertension. Although iNO can be given via nasal canula, sildenafil can be very useful in this situation.

Dose of sildenafil: Starting dose 0.1 to 0.2 mg/kg/dose every 6-8 hours. Can be increased to 1 to 1.5 mg/kg/dose every 6 hours. Possible side effect includes hypotension requiring fluid bolus or inotropes.

**Volume and inotropes**

1. There is no evidence to suggest the routine use of inotropes or fluid boluses in pulmonary hypertension in CDH.

2. The aim should be to keep the infant normotensive. For a term baby mean blood pressures of 40 mmHg are acceptable.

3. In the presence of low systemic blood pressures, 1-2 fluid boluses can be given before inotropes are considered.

4. Milrinone can be used as a first line inotrope as it can improve right ventricular output and is less chronotropic than other inotropes. Other inotropes that can be used are dopamine and dobutamine and rarely epinephrine.

5. The routine use of steroids to improve systemic blood pressure is not recommended. However, if hypotension is resistant to inotropes and fluid boluses and other causes of hypotension such as pneumothorax and sepsis are ruled out, hydrocortisone may be considered after drawing cortisol levels.

**Non-pharmacological interventions**

Although there is no scientific evidence, the importance of non-pharmacological interventions cannot be over emphasized. This would include things like minimal handling, a quiet environment and alleviation of pain and discomfort.

**Transfer of patients to ACH**

1. Prior to transfer, a multidisciplinary meeting involving neonatology, cardiology, PICU, surgery and preferably anesthesia should take place.

2. The infant should demonstrate a period of stability for 48 h prior to transfer. Optimally the infant should be on a FiO₂ ≤ 0.4, MAP of ≤ 12 cmH₂O and preferably be off inotropes and pulmonary vasodilators. However, these criteria could vary depending upon the infant’s clinical course. If the infant is on higher ventilator settings and is also on pulmonary vasodilators and/ or inotropes but has been stable for 48 h and it is felt that no further improvement is likely, transfer at settings higher than mentioned above can be considered. **This, however, must be discussed with cardiology, surgery and PICU prior to transfer of the baby.**

3. The need for an ECHO prior to transfer to ACH should be discussed with cardiology
The Role of ECMO

The role of Extra Corporeal Membrane Oxygenation (ECMO) in the management of the neonate with CDH remains controversial.

In the US, the use of ECMO in CDH patients varies from 11% - 61% of patients. Some centres have reported a reduced requirement for ECMO and also an improved survival with ECMO in CDH in the era of “gentle ventilation” management techniques.

In Canada, the use of ECMO for patients with CDH is uncommon, with many centres reporting poor survival with ECMO in this population.

There are ECMO programs in Edmonton and Vancouver.

The Edmonton program has not successfully used ECMO for a patient with CDH in over 5 years and does not recommend its use in this group of patients.

The Vancouver program is currently reviewing their practice in light of low survival rates with ECMO in CDH patients.

Although still commonly used in US centres, ECMO for neonatal CDH patients is currently not available here in Calgary and is not recommended by the provincial ECMO program in Edmonton.

In the past, some antenatal patients, whose fetus with CDH had antenatal markers associated with a higher mortality risk, were encouraged to deliver in Edmonton because of the availability of ECMO in that centre.

At the present time, because of above recommendation from the Edmonton ECMO program, there is no advantage for the baby to be delivered in Edmonton - the exception being a baby with a serious cardiac lesion in addition to the CDH.

Following extensive multidisciplinary discussion, (with representatives from Maternal Fetal Medicine, Obstetrics, Neonatology and Pediatric Surgery) we concluded that, currently, ECMO has no role in the management of our patients with CDH.

The addition of this consensus (re the role of ECMO) to the CDH management guideline will improve the consistency of the care we can offer to the CDH patients. It will also avoid unnecessary patient transfers either of the mother antenatally or the baby postnatally.

CNS CONSIDERATIONS

The overall goal is to minimize the infant’s stress and discomfort and thereby minimize exacerbation of the underlying pulmonary hypertension.

Environment

1. NICU Bed: determine ahead of the delivery where the infant with CDH will be admitted. Optimally a spot should be chosen that may be completely sectioned off or at least has minimal traffic and extraneous noise such as telephones/intercoms, other busy/noisy patients.

2. BED Type: the OmniBed should be used; the lid should be in the down position as much as possible.
3. Alarms: limit the number and volume of alarms. If the infant is in a quiet part of the nursery with one to one nursing, the need to have alarm volumes set at maximum should be eliminated.

4. Limit Conversations: foster an environment of reduced noise and calm at every opportunity. This should include performing ward rounds away from the bedside and never talking over the infant him/herself.

5. Light Levels: when lighting increased for interventions, the infant’s eyes should be protected with a cover.

**Bedside Interventions**

1. Simple comfort measures should be tried prior to pharmaceutical interventions:
   - Provide developmentally appropriate positioning (midline positioning, flexion, containment)
   - Positioning aids may be of use e.g. bean bags to keep ventilator tubing in place and limit pulling.
   - Provide two or more person care-giving for procedures or interventions.
   - Handling should be gentle and without sudden changes.
   - Consider giving a soother for non-nutritive sucking +/- 24% sucrose.
   - Cover the eyes to limit exposure to bright light.

2. Limit the number of interventions and maximize automated monitoring.

3. Prior to any significant intervention, such as blood letting, ETT suctioning, turning the infant, ensure adequate sedation and analgesia.

4. **Maintaining stomach and bowel decompression is essential.** Gaseous distention of the herniated bowel frequently causes destabilization in respiratory and cardiovascular status.
   - Reassess patency of the Replogle on an hourly basis.
   - Irrigate the Replogle tube with 2 mls of warmed normal saline every 4 hours.
   - Standard practice is to start Replogle suction pressure at 20 mmHg.
   - To obtain and maintain optimal bowel/stomach decompression the medical team may order the suction pressure to go as high as 50 mmHg.
   - If increasing suction pressure is necessary this should be done in 10 mmHg increments.

5. Assessments and interventions should be clustered as much as possible in terms of number and duration if the infant can tolerate the handling.

6. Painful procedures should not be performed at the same time as other, non-emergency routine care (e.g., taking vital signs, changing a diaper).

7. Ensure the infant can be visually observed at all times.

8. X rays should be taken using the frame under the OmniBed.

9. Weighing should be performed by the OmniBed weighing scale.
Sedation and analgesia

In general, agitation and discomfort are to be avoided if at all possible. However, over sedation is associated with reduced respiratory effort which may be detrimental in this population. Sufficient sedation and analgesia are required to minimize perturbations in the pulmonary vascular resistance but should also allow for spontaneous respirations and some movement.

Opioids

All opioids have the propensity to cause respiratory depression, hypotension, gut hypomotility and urinary retention. These should be monitored and managed accordingly.

Fentanyl:
- This agent may have some pulmonary hypertension relieving properties and therefore should be first line. It is also more stable hemodynamically than morphine.
- Starting dose is 1-2 \( \mu \)g/kg/hour
- Intravenous bolus doses should be given slowly over 5 minutes, minimum, to limit the risk of developing chest wall rigidity.
- Tachyphylaxis occurs quickly and the dose will need to be increased at least daily
- The maximum safe dose is unknown but a reasonable range is 2-10 \( \mu \)g/kg/hour
- Anticipate withdrawal symptoms if fentanyl is used for 5 days or more and is reduced abruptly.

Morphine:
- Morphine has less cardiovascular stability than fentanyl.
- If fentanyl seems insufficient, it is reasonable to consider morphine as a second line agent.
- Starting dose will depend on the timing of its introduction but there is no upper limit. In general, starting morphine at 10 times the dose of fentanyl used will be sufficient.
- Monitor carefully of blood pressure is required. Rarely bronchospasm is an issue.

Benzodiazepines

Benzodiazepines are well known anxiolytics, producing sedation and muscle relaxation thereby improving synchrony with assisted ventilation in neonates. Although they may lessen the opiate dose required, they do not provide analgesia. Side effects include respiratory depression, hypotension especially if the infant has an opioid infusion running concurrently, and occasional neuronal excitability in the form of myoclonic jerks. These should be monitored and managed accordingly.

Of significant concern are reports of excessive neuronal apoptosis in the brains of newborn animals given benzodiazepines. Although extrapolation to the human may not be appropriate necessarily, the use of benzodiazepines should be minimized and the risks acknowledged.

Midazolam:
- A short acting benzodiazepine, but individual responses vary greatly
- Caution required if the infant is hemodynamically unstable as midazolam is a vasodilator and may cause a drop in blood pressure.
- Usually used in conjunction with an opioid.
- Starting dose is 30 \( \mu \)g/kg/hour. Maximum dose is unknown but may be over 300 \( \mu \)g/kg/hour.
Other Sedatives

Phenobarbital:
Phenobarbital is frequently used to potentiate the effect of analgesics such as morphine or fentanyl. It has little to no analgesic effect itself. Side effects are not dissimilar to those of opiates with respiratory depression, hypotension, tolerance and dependence. As there is little to prove its efficacy in the neonatal population and there are concerns about its long term effect on the CNS, its use is not recommended.

Neuromuscular Blocking Agents (NMBA)

In this modern era of permissive hypercarbia, the use of NMBA is strongly discouraged. However, in the event of uncontrollable clinical deterioration and/or inability to achieve adequate pre-ductal oxygen saturation or ventilation, there may be a need for temporary neuromuscular blockade. In that instance, the shortest possible time without spontaneous movement and respirations should be the goal.

Rocuronium
One of the newer intermediate duration acting NMBA, rocuronium has the advantages of rapid onset and impressive cardiovascular stability, with no histamine release, even with large doses. It should be the first line agent in our population. The pharmacokinetics of rocuronium are altered by renal and hepatic disease; the latter probably has the more significant effect.

- The IV bolus dose is 0.5mg/kg/dose every 20 to 30 minutes. It may be run as an intravenous infusion at 10-20mcg/kg/minute.

Vecuronium
Vecuronium is another intermediate duration acting NMBA with good cardiovascular stability.

- The IV bolus dose is 0.1mg/kg/dose

Pancuronium
Most NICUs are very familiar with pancuronium. However, it can cause tachycardia, increased blood pressure, and increased cardiac output. Also, hypotension and decreased oxygenation acutely, and muscle weakness and contractures with longer term use, have been reported.

- The dose is 100mcg/kg/dose, repeated every 30-60 minutes as required.

Transport of CDH Patient to ACH

- Every effort will be made to transport the baby with CDH only when stable (respiratory and cardiovascular support unchanged for the previous 24-48 hours).
- Transport timing will be coordinated with the multidisciplinary team.
- PICU Attending contacted and case is discussed. Surgeon should also be made aware and supportive of the baby’s transfer.
- A Neonatologist will, in most instances, be available to accompany the high risk neonatal transport team when moving a baby with CDH.
- In addition to standard neonatal transport care, special attention may be required for the following systems:
  - Analgesia/sedation
The infant will have been maintained on minimum analgesia/sedation in the NICU. A bolus of fentanyl and increased infusion rate will most likely be required for transport.

Minimal handling and stimulation will continue to be important (efficient moving, supportive nesting, ear muffs).

- **Respiratory**
  - At this time, the transport ventilator only supports a conventional mode. iNO may be administered.
  - Once the baby has been stable on the transport ventilator for 5-10 minutes an arterial blood gas should be checked. An ABG should also be documented upon arrival at the receiving hospital.
  - Avoid manual bagging ventilation if possible. Target Sp02/TcPc02 to be ordered by responsible Neonatologist.
  - Continual decompression of the stomach is critical to maintaining stability.

- **Cardiovascular**
  - Cardiac support drugs should be transitioned to transport syringe pumps at least 1 hour prior to estimated time of departure.

- **Other**
  - Lipids may be held for the transport.
  - Family will be aware of estimated departure time.

**OUTCOME & FOLLOW-UP**

Increase in survival in CDH patients often comes with greater morbidity such as cardiopulmonary, gastro-intestinal and neurological problems in approximately 87% of CDH survivors. Associated cardiac or chromosomal anomalies in children with CDH may involve a wide range of problems, which need multidisciplinary follow-up.

**Beginning January 1, 2011 all surviving CDH patients will be followed at a multidisciplinary clinic at ACH**

**Pulmonary morbidity**

Long Term Pulmonary sequelae in 30–50%.
Lung function abnormalities in 28–52%.
Chest X rays abnormalities in 33–80%.

**Cardiovascular morbidity**

Pulmonary vascular bed abnormality.
Altered expression of factors involved in regulation of vascular tone.
Pulmonary hypertension.
Hypoperfusion on the affected side.
Long-term consequences are unknown.

**Gastro-intestinal morbidity**

Gastroesophageal reflux (GER) up to 84% and 23% of those will require surgery.
Intestinal obstruction in up to 20% of CDH survivors. Recurrence rate (ie, re-herniation) in up to 40%, but lately between 6% to 7% range.
Failure to thrive related to GER, catabolic stress, oral aversion and increased caloric requirement due to pulmonary morbidity.

Up to 30% of the patients remain below the 5th percentile for weight, despite optimal caloric intake.

Infants requiring ECMO because of CDH are at greater risk for lower weight, length and Weight/Length ratio

Routine nutritional assessment with emphasis on weight, length, weight/length ratio, growth velocity and head circumference with early nutritional intervention is essential to optimize growth in these patients.

**Neurological morbidity**

Neurodevelopmental problems are common with sensorineural hearing loss up to 10% at the age of 3 years, motor problems in 60% during the first year and in 73% at the age of 3. Language problems can be seen in 60% of the children at the age of 3 years.

Social and behavioral problems occur in approximately 10% of the cases. A mean IQ of 85 has been documented and only half of these children were at the expected school level.

Long-term follow-up is important in CDH survivors. The nature of the multisystemic long term complications makes it imperative that these infants receive comprehensive follow up by a multidisciplinary team.

Regular neurological, developmental and (neuro) psychological assessment by a specialized pediatrician and a child’s psychologist is recommended. In case of motor, cognitive, speech and behavioral problems, further treatment by a physiotherapist, a speech therapist or a child’s psychologist needs to be considered. Therefore, neurodevelopmental follow-up is preferably in the hands of a multidisciplinary team.

That team should consist of neurodevelopmental specialist, surgeon, gastroenterologist, and pulmonologist, cardiologist, and ancillary services such as language therapist, occupational therapist.

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