Management of Congenital Diaphragmatic Hernia in Infants

Procedure Responsibilities and Authorisation

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Procedure Review History

Version	Updated by	Date Updated	Description of Changes
3	Dr David Bourchier	25/2/16	Changes to the values in the progress graph

Management of Congenital Diaphragmatic Hernia in Infants

A. Purpose of procedure:

Management of congenital diaphragmatic hernia

B. Background:

Congenital Diaphragmatic Hernia (CDH) has an incidence of approximately in 1 in 3000 live births. 85% are left sided. There could be a 40 – 50% association with other malformations. Survival depends on the degree of hypoplasia of the lungs as well as other associated lethal anomalies[1]. Pulmonary hypertension of varying severity is invariably present and is also an important determinant of survival[2]. Despite several advances in the ventilatory strategies, including the use of nitric oxide, the outcome has remained stubbornly unchanged with a survival rate of around 60 – 70%. Several centres from the developed world have started reporting survival statistics up to 80 – 90% using a protocol-based management [3,4,5,6]. The key principle of this protocol is strict avoidance of high airway pressure ventilatory management from birth, acceptance of low (down to 85%) pre-ductal saturation and permissive hypercapnia. Barotrauma has now been recognised as a significant cause of mortality and morbidity in CDH[7]. This could be avoided by using this strict protocol-based management, thus hopefully improving the survival statistics.

Of late, some centres around the world are reporting better survivals with the use of ECMO in severe CDH (about 10% of CDH would fall into this group)[8]. As such, an early referral to PICU at Starship should be considered. Ideally engaging with them before the birth of the baby is advised, with in utero transfer as an option for the family.

C. Management at Birth:

- Every known case of CDH should have in attendance neonatal personnel capable of intubation.
- Avoid bag and mask ventilation and CPAP.
- Electively intubate – **avoid hyperinflation at all times by restricting PIP to 25 cms or less**.
- Decompress the stomach with NG tube.

D. Management in NICU:

- Sedate lightly, allowing the babies to breathe spontaneously.
- Use paralysis sparingly and only if the infant is struggling against the ventilator.
- Initial ventilatory setting: PIP below 25 cms of water, PEEP 4-5 cm and rate 40 -60/min, FiO2 enough to maintain pre-ductal saturations ≥ 85%.
- Insert UAC/ PAL (on the right radial preferably) and UVC/ CVL
- Monitor pre-ductal saturations (Sats monitor or PAL on right arm)
- Perform cardiac ECHO to rule out CHD, to assess the degree of pulmonary hypertension and right ventricular function.
- Keep the High Frequency Oscillator (HFO) and nitric oxide on standby.

E. Goals of Management:

(Use the attached CDH: Progress-graph for charting blood gas results and ventilatory parameter)

- Avoid high airway pressures. Switch to HFO if the PIP on conventional ventilation cannot be maintained below 25 cms water.
- When using HFO keep the MAP below 16 cm. Start with a frequency of 10 Hz.
- Switch back to conventional ventilator when stable and ABGs show improvement.
- Tolerate pre-ductal saturation down to 85%. *Do not make ventilatory changes based on post-ductal saturation or post-ductal ABG*. (Always keep the consultant informed before any major change)
- Tolerate moderate hypercarbia of up to 8 kPa, as long as the pH is over 7.2

- Avoid hypocarbia and alkalosis.
- Correct metabolic acidosis (pH below 7.2) by bicarbonate infusions and/ or volume replacement.
- Use nitric oxide when the pulmonary arterial pressure equals, or exceeds, the systolic blood pressure, as evidenced by right to left ductal shunting on ECHO
- Maintain the systolic BP over 50 mm of Hg, if necessary by volume-loading and/or inotropes
  (dopamine, noradrenaline) or hydrocortisone.

F. Surgery:

When the infant is stable on the following parameters for 24 – 48 hours.
(No advantage in unduly delaying surgery – it may cause further lung damage)

- The baby on conventional ventilator with PIP below or equal to 25.
- The oxygen requirement is low- about 50%.
- The inotropic support is minimal.
- Nitric oxide is off.
- The pulmonary artery pressure is roughly 50% of systolic blood pressure.

G. Post-operative management:

- Same principles as pre-operative management.
- Start enteral feeds as soon as the post-op ileus has resolved.

H. References:

1. Langham MR Jr et al. CDH: Epidemiology and Outcome. Clin Perinatol 1996; 23:671 – 688.
2. Dillon PW et al. The relationship of pulmonary artery pressure and survival in CDH. J Pediatr Surg 2004;p 39: 307 – 312.
3. Boloker J et al. CDH in 120 infants treated consecutively with permissive hypercapnea/spontaneous respiration/elective repair. J Pediatr Surg 2002; 37:357 – 366.
4. Downard CD et al. Analysis of improved survival rate for CDH. J Pediatr Surg 2003; 38(5):729 – 732.
5. Bagola p et al. Impact of a current treatment protocol on outcome of High-Risk CDH. J Pediatr Surg 2004; 39(3):313 -318.
6. Beca J. Outcome statistics using protocol based management in CDH. PICU Starship Children's Hospital, Auckland. Personal Communication 2007.
7. Bohn D. Congenital Diaphragmatic Hernia. Am J Respir Crit Care Med. 2002; 166:911 – 915.
8. Kays D. Wéal. Outcomes in the physiologically most severe CDH patients. J. Pediatr. Surg 2015, 50:893—97.

Congenital Diaphragmatic Hernia: Progress-graph

Date _____

	7.6												
	7.4												
pH	7.2												
	7.0												
	6.8												
	16.2												
	13.3												
pCO2	10.8												
	8.1												
	5.4												
	2.7												
PIP	40												
	30												
	20												
HFV MAP	25												
	15												
	5												
		0	2	4	6	8	10	12	14	16	18	20	22	24

Hour of Day