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9-2-2009

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Recommended Citation
Weitzel, Brett; Gerald Holmes; and Rebecca Moran. "The Modified McGoon Index does not Predict Mortality in Infants with Congenital Diaphragmatic Hernia." (2009). https://digitalrepository.unm.edu/ume-research-papers/95

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The Modified McGoon Index does not Predict Mortality in Infants with Congenital Diaphragmatic Hernia

Brett Weitzel, Gerald Holmes, MD, Rebecca Moran, MD

Abstract

Background: Infants diagnosed with congenital diaphragmatic hernia (CDH) continue to have a high mortality despite medical advances such as inhaled nitric oxide (iNO), high frequency ventilation (HFV) and/or extracorporeal membrane oxygenation (ECMO). The mortality in some studies exceeds 80%. The modified McGoon index (MMI), a ratio of pulmonary artery size to aortic size has been reported to predict survivability in these infants: infants with a MMI ≤ 1.3 have an 85% mortality: (*Pediatr* 2000;105:1106).
Objective: To evaluate the utility of MMI in predicting mortality in our population.
Design/Methods: We performed a retrospective review of all infants with CDH admitted to the University of New Mexico NICU between January 2000 and November 2002. All available echocardiograms were reviewed by a single pediatric cardiologist blinded to the outcome of the infants, and the MMI was calculated (LPA + RPA/ Ao). The following information was also recorded from each infant's record: gestational age, birth weight, Apgar scores, need for cardiopulmonary resuscitation in the delivery room, length of hospital stay, days of ventilation, use of high frequency ventilation, use of iNO, use of ECMO and survival to discharge.
Results: Eighteen infants with CDH were admitted; echocardiograms were available for 15. MMI ranged from .06 to 2.37 in the 15 infants. Seven infants died and 8 survived to discharge. There was no significant difference in birth weight between groups (lived (L): 3151 ± 203; died (D): 2735 ± 186 grams, mean ± SE), gestational age (L: 38.3 ± .4; D: 38.3 ± 1.1), CPR in delivery room (L: 1/8; D: 3/7). Infants who died had lower 5-minute Apgar scores (L: 7.1± .5; D: 4.8 ± .7, $p < 0.05$). A greater number of infants who died received ECMO. There were no differences between groups in the number of infants on HFV or iNO. An MMI of ≤ 1.3 did not predict death in our patient population: mortality in patients with an MMI ≤ 1.3 was 46% (6 of 13 infants), while mortality in patients with MMI ≥ 1.3 was 50% (1 of 2 infants).
Conclusion: MMI ≤ 1.3 did not predict mortality in our population.

Background

Congenital diaphragmatic hernia (CDH) refers to a developmental abnormality in the diaphragm that allows abdominal viscera to herniate into the chest during the critical period of fetal lung development when the bronchi and pulmonary arteries are undergoing branching. CDH has an incidence of approximately one in 2,500 live births[1-3] and is associated with significant morbidity and mortality rates. Approximately 80% of these irregularities are found on the left side of the diaphragm. The most common form is the classic posterolateral, also known as Bochdalek, hernia. CDH occurs in conjunction with other anomalies 40-50% of the time[4]. The central nervous system is the most commonly affected system. Congenital heart malformations are also associated with CDHs, and are the most important comorbidities when considering prognosis. CDH is also associated with chromosomal abnormalities, including Turner's syndrome, and trisomy 13 and 18. Chromosomal irregularities are also associated, such as Fryn's Syndrome. Familial associations have been reported although they are very rare[5].

Despite medical advances in treatment methods, including inhaled nitric oxide (iNO), high frequency oscillation ventilation (HFOV), gentle ventilation with permissive hypercapnea, delayed surgical repair and extracorporeal oxygenation (ECMO), mortality rates continue to be high. Some studies have shown mortality rates greater than or equal to 80%[6], although they reflect the outcomes of fetuses with life threatening chromosomal and anatomic anomalies as well as those with isolated CDH. Commonly quoted survival statistics for isolated CDH are 80% for those infants who do not require ECMO and 50% for those who do.

Antenatal diagnosis of CDH has been possible for more than two decades and currently prenatal ultrasounds are used to diagnosis almost 50% of infants with CDH early in the pregnancy at 16-24 weeks of gestation. The most common findings on ultrasound are the presence of the stomach in the left chest, which causes the mediastinum to shift to the right.

Aside from the coexistence of chromosomal abnormalities or congenital malformations, no antenatal findings have been able to accurately predict death in infants with isolated CDH. Studies in the 1980's demonstrated an increased mortality with diagnosis of CDH early in gestation[7,8]. But with the advent of high-resolution ultrasonography and growing experience in the prenatal diagnosis of CDH, earlier diagnosis of less severe disease has become more common. In the 1990's additional features seen on antenatal sonograms were proposed as predictors of CDH severity. The findings that were studied included polyhydramnios[9,10], thoracic stomach bubble[11], mediastinal shift[9], lung-thoracic transverse area ratio[12,13], left ventricular disproportion before 25 weeks gestation[14], right lung area to head circumference ratio[15], contralateral lung area and fetal branch pulmonary artery diameters[16]. All of these proposed predictors have failed to be substantiated in other studies.

The establishment of predictors of postnatal morbidity and mortality due to CDH are important in antenatal counseling and planning of perinatal treatment. These predictors may also impact perinatal palliative care planning in the case of a lethal lesion. In 2001 Suda et al proposed the Modified McGoon Index (MMI) as a CDH severity predictor[17]. The MMI is calculated as the combined diameter of hilar pulmonary arteries, indexed to the descending aorta at the level of the diaphragm. Their data showed that the MMI had both a high sensitivity and specificity for mortality due to CDH. Their findings were corroborated in 2006 by Casaccia et al[18], but disputed by Yao et al[19] in 2004. In the Suda study, a MMI less than 1.3 predicted mortality with a sensitivity of 80% and a specificity of 100%[17].

Introduction

The aim of our study was to evaluate the utility of the MMI in predicting mortality in our population. We also sought to determine whether antenatal MMI ratios and other parameters correlate with postnatal outcome and can be used to stratify CDH into mild and severe cases.

Methods

We performed a retrospective review of all infants with congenital diaphragmatic hernia admitted to the University of New Mexico Neonatal Intensive Care Unit between January 2000 and November 2002. All available echocardiograms were reviewed by a single pediatric cardiologist blinded to the outcome of the infants, and the modified McGoon Index was calculated (LPA + RPA/ Ao). The following information was also recorded from each

infant's record: gestational age, birth weight, Apgar scores, need for cardiopulmonary resuscitation in the delivery room, length of hospital stay, days of ventilation, use of high frequency ventilation, use of iNO, use of ECMO and survival to discharge.

Results

Eighteen infants with CDH were admitted during the 23 months studied; echocardiograms were available for fifteen. MMI ranged from .06 to 2.37 in the 15 infants. Seven infants died and 8 survived to discharge. There was no significant difference in birth weight between groups (survived (S): 3151 ± 203; died (D): 2735 ± 186 grams, mean ± SE), gestational age (S: 38.3 ± .4; D: 38.3 ± 1.1), or CPR in delivery room (S: 1/8; D: 3/7). Infants who died had lower 5-minute Apgar scores (S: 7.1± .5; D: 4.8 ± .7, $p< 0.05$). None of the infants who survived received ECMO during the study period. There were no differences between groups in the number of infants on HFV or iNO. An MMI of ≤ 1.3 did not predict death in our patient population: mortality in patients with an MMI ≤ 1.3 was 46% (6 of 13 infants), while mortality in patients with MMI ≥ 1.3 was 50% (1 of 2 infants).

Conclusion

This small retrospective study did not support Dr. Suda's findings of an MMI less than 1.3 predicting mortality[17]. As others have reported, the need for ECMO negatively impacts survival. In our study population, one infant had chromosomal derangements. In this infant a 6:12 rearrangement was discovered. This infant was also born premature at 34 weeks gestation and never underwent surgical repair of the lesion. This infant died at 23 days of life. There were four additional infants who were not repaired: 2 were premature infants, of which one had congenital heart disease. Another infant of the unrepaired group experienced cardio-pulmonary arrest during ECMO cannulation and the other had extension of a intracranial hemorrhage on ECMO and was withdrawn from support prior to surgical repair of the hernia.

The limitations of the study include the retrospective nature of the study design. After the release of the Suda article our institution was eager to determine if this measurement was applicable in our population and was a means for predicting appropriate candidates for aggressive management and consideration of ECMO. As CDH is a relatively rare diagnosis the most appropriate initial investigation was a retrospective review to determine if we should then proceed with prospective data collection.

In summary, the MMI in our small retrospective review of 15 infants with CDH did not correlate with mortality. We need to aggressively evaluate methods for antenatal and immediate post-natal prediction of survival to aid in delivery planning, palliative care plans when appropriate and prognostication for these critically ill children and their families.

Acknowledgement

We would like to thank Robin Ohls for her assistance with the statistical analysis of this paper.

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