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Perioperative course of pulmonary hypertension in infants with congenital diaphragmatic hernia: Impact on outcome following successful repair

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Background: Pulmonary hypertension (PH) associated with congenital diaphragmatic hernia (CDH) remains a significant cause of morbidity and mortality. For improved outcomes, preoperative stabilization is commonly practiced to control the PH in infants with CDH. Some CDH infants who have been considered stabilized and ready for surgery have nevertheless developed significant PH after surgical repair. In fact, the markers and consequences of the preoperative stabilization are still unclear. Therefore we examine the peri-operative course of PH to evaluate the impact of preoperative PH severity on mortality and morbidity of infants who underwent surgical repair of CDH.

Methods: The medical charts of all newborns ($n = 49$) with CDH who were treated at our institution between January 2000 and December 2009 were reviewed. General management and perioperative data were evaluated for all infants. The ratio of estimated pulmonary artery pressure to systemic pressure (P/S ratio or PSR), based on echocardiographic data, was used to assess the PH severity during the perioperative period.

Results: The overall survival rate in our group of infants with CDH was 71.4%. Of the 49 infants with CDH, 9 (18.4%) died during the preoperative phase. Forty infants underwent CDH repair at a median age of 3.5 days (range, 1 to 46 days). Five of these infants (12.5%) subsequently deteriorated and died after surgery. Using ROC analysis, a PSR cut-off value prior to surgery of 0.9 predicted mortality in CDH infants, with a sensitivity of 100% and specificity of 84% and with an area under the curve (AUC) of 0.93 ($P=0.002$). Accordingly, two groups of infants with distinct outcomes were identified, as follows: a low-PSR cohort ($PSR \leq 0.9$) with a survival rate of 100% and a high-PSR cohort ($PSR > 0.9$) with a survival rate of 50% ($P=0.001$). The rate of pneumothorax and the frequency of use of several inotropic agents after surgery were significantly higher in the high-PSR group ($P=0.001$ and 0.007 , respectively). Compared with low-PSR infants, infants with high PSR were operated on later ($P=0.03$) and were postoperatively ventilated for longer ($P=0.01$). During the entire perioperative period, significant differences in the PH severity were noted between the two PSR groups. During the first week of life, infants in the high-PSR group had significantly higher PSRs than those in the low-PSR group ($P=0.001$), and similar tendencies continued to be significant between the two groups after CDH repair ($P=0.04$). **Conclusions:** During the perioperative period, PH severity monitoring via the serial assessment of PSR is beneficial. Better outcomes were observed with a preoperative $PSR \leq 0.9$, and this association needs to be confirmed by prospective study.