TIMING TO PGE INITIATION AND THE NEED FOR ADDITIONAL PH THERAPY: A SINGLE CENTER EXPERIENCE

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Background: Congenital diaphragmatic hernia (CDH) is a congenital anomaly in which a diaphragmatic defect allows herniation of abdominal contents into the thorax, leading to pulmonary hypoplasia and pulmonary hypertension (PH) requiring surgical repair. Despite advances in management, mortality rates remain 20%–30%, with PH as a major determinant of survival and outcome. Prostaglandin E1 (PGE) has been used in neonates with CDH and severe PH to maintain ductus arteriosus patency, thus relieving the pressure-loaded right ventricle and augmenting systemic blood flow. We aim to characterize how timing of PGE relates to the need for additional pulmonary vasodilators in infants with CDH at a large, tertiary care center.

Materials/Methods: We performed a retrospective cohort study of patients followed by the TCH Pulmonary Hypertension team from January, 2011 through September, 2021. Alprostadil exposure in the NICU was queried, and patients cross-referenced to an existing CDH database to ensure completeness. Patients with suspected or confirmed cyanotic congenital heart disease, coarctation of aorta, or additional major structural birth defects were excluded. We compared demographics, clinical characteristics, and timing of PH therapies, stratified by prenatal markers of disease severity. Bivariate and descriptive statistics were used to determine clinical characteristics and outcomes.

Results: We identified n=18 (gestational age 37w2d s=2d) inborn infants with an isolated prenatal diagnosis of CDH who were exposed to PGE during the study period. 12/18 (67%) were male and average birth weight 2883 g (s=702 g). 13/18 (72%) had left CDH, 1/18 (6%) bilateral, and 4/18 (22%) right. Observed to expected total fetal lung volume (O:E TFLV) was 0.28 (s=0.129) with a percent liver herniation of 25.8% (s=12.4). 15/18 (83%) infants were classified by prenatal O:E TFLV as having severe CDH. Overall mortality rate for the cohort was 33% with 12/18 infants surviving to discharge. 16/18 (94%) of infants were exposed to sildenafil at any point during initial hospitalization, and 8/12 (67%) surviving infants were discharged home on sildenafil.

Conclusions: Infants exposed to PGE at our institution had severe markers of prenatal disease, and all 18 infants were exposed to inhaled nitric oxide (iNO) for treatment of pulmonary hypertension prior to repair. Of the 12 surviving infants exposed to iNO, sildenafil and PGE combined, 3 were able to be weaned off oral therapy prior to discharge despite severe prenatal markers of disease.

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