

Pulmonary Disorders 2017: Respiratory management of the newborn with an omphalocele_Joanne Baerg_Loma Linda University Children's Hospital, USA

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An omphalocele is a congenital defect of the umbilical ring with herniation of the viscera. Despite of advances in neonatal care, for live-born infants, the mortality rate remains between five and 25%. Respiratory failure at birth is an independent predictor of mortality for omphalocele infants, but the causes are diverse. In this presentation, giant and non-giant omphalocele are compared, as giant omphalocele have more respiratory difficulties. Prenatal predictors of post-natal respiratory failure and care strategies are discussed. Pulmonary hypoplasia is defined. Historically, fetuses and infants with omphalocele are reported to have markedly reduced chest capacities. Recently, fetal magnetic resonance imaging (MRI) has expanded the understanding of decreased congenital lung volume in infants with omphalocele. Clinical-radiologic correlation studies support the use of prenatal MRI to predict the degree of respiratory insufficiency observed in the postnatal period. The contribution of major abnormalities to breathing difficulties is discussed. Infants with omphalocele may have increased pulmonary vascular reactivity and pulmonary hypertension which increases the risk of postnatal mortality. In this presentation, pulmonary hypoplasia and pulmonary hypertension are defined as separate entities. The two diagnoses must be distinguished from each other in the clinical setting. The implications of congenital heart defects are explained. The role and objectives of assisted ventilation in respiratory failure are expanded. Since 2011, infants with omphalocele and respiratory failure have required the extracorporeal membrane oxygenator. The first review of the Extra-Corporeal Life Support (ELSO, Ann Arbor, MI USA) database for causes of respiratory failure and outcomes in omphalocele infants will be presented. The timing of surgical repair, postoperative complications such as compartment

syndrome, delayed surgical closure techniques, and the implications of a ruptured omphalocele are explained. Abnormalities in lung function, chronic lung disease, the role of tracheostomy, influence of gastroesophageal reflux disease (GERD), prematurity, and improved outcome strategies are discussed.

The giant omphalocele represents an important subset of omphalocele infants. Giant omphalocele is defined as an omphalocele defect containing more than 75% liver in the sac and / or a diameter greater than 5 cm. A non-giant omphalocele is generally defined as a defect with a diameter of less than 5 cm. Previous definitions that measured the defect in centimeters do not take into account differences in height and gestational age of infants.

The definition that giant omphalocele is a defect containing more than 75% liver in the sac is preferable and consistent. Giant omphalocele is associated with a poor prognosis in many studies. Giant omphalocele is often associated with a higher incidence of respiratory failure, longer ventilation requirements, and an increased incidence of pulmonary hypoplasia and pulmonary hypertension.

Infants with giant omphaloceles have significantly higher neonatal morbidity. When large and small omphaloceles are compared, median length of stay (47 vs 10 days), median age at complete enteral feeding (23 vs 5 days), median duration of mechanical ventilation (23 vs 7 days) and supplemental oxygen requirements at 30 days of life (88% vs. 27%) are significantly longer for infants with giant omphaloceles. Respiratory failure is the leading cause of death in infants with giant omphaloceles. In the neonatal period, these infants have significantly more pulmonary hypoplasia and pulmonary hypertension and therefore more breathing difficulties