A Concise Message on CDH: Rare—Yes; Risky—Probably

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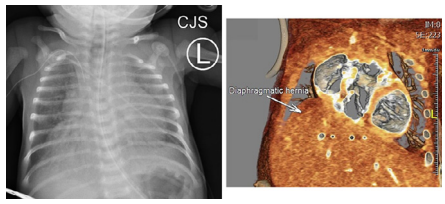
Fraser et al from the Johns Hopkins Hospital present an analysis of the prevalence and impact of congenital diaphragmatic hernia (CDH) in patients undergoing congenital heart surgery. They utilize data from the Society of Thoracic Surgeons Congenital Heart Surgery Database spanning 7 years, from 2010 to 2016. Among more than 150,000 patients, CDH was coded in only 426, or just under one-third of 1%. The authors found that CDH was associated with increased operative mortality, complications, and length of stay, particularly in neonates and infants.

In the past, the relationship between CDH and congenital heart disease has been examined from the CDH perspective. We know that CDH is rare, with an incidence of about 1 in 3000 births.[1] Among babies with CDH, from 10% to 35% also have a congenital heart defect.[2] The additional presence of a heart defect increases CDH mortality by about 30%.[2] The outcome for CDH with a single ventricle heart defect is especially poor, with survival as low as 5%. The current study comes at this relationship from the opposite perspective, which is the first time this has been done in a comprehensive fashion. Among children undergoing surgery for congenital heart disease, the frequency of CDH is very low (0.27%), but is still 5–10 times the prevalence in the general population, reinforcing the fact that congenital heart disease and CDH are associated with each other. The observed morbidity and mortality accompanying a diagnosis of CDH in neonates and infants is quite pronounced. Among neonates with CDH undergoing STAT 3–5 operations, mortality was 34%, length of stay was almost 2 months, and postoperative complications occurred in 90%. While perhaps not surprising, these findings are certainly striking.

The primary limitation of this study is the lack of information on the details of the CDH. The outcome of any patient with CDH is affected by the severity of the diaphragmatic defect, the need for and timing of surgical repair, as well as associated pulmonary hypoplasia and pulmonary hypertension. These variables, in turn, affect the need for Extracorporeal membrane oxygenation (ECMO), which is utilized in 30–40% of CDH patients.[1,3] In its current iteration, the STS database codes only for the presence of CDH (yes or no), but not for any of these other factors. From this data, we cannot tell whether the CDH was repaired, and if so, whether it was repaired before or after the cardiac operation. These limitations make it difficult to apply the current study's findings to individual patients.



Images from a neonate with congenital heart disease and a right-sided diaphragmatic defect.

Central Message

In patients undergoing congenital heart surgery, a congenital diaphragmatic hernia occurs in 0.27%, and appears to increase operative risk.

It should also be noted that the CDH and non-CDH groups were not equivalent, with the CDH patients having a higher incidence of a chromosomal abnormality or syndrome, and more frequent preoperative mechanical ventilation. It seems intuitive that CDH is an independent risk factor for poor outcome, but the relative impact compared to other risk factors is unknown. While the authors stratified their patients by age and primary procedure, they did not incorporate the other variables available in the STS Database mortality risk models. Although the number of patients may have been too small to allow a formal multivariable analysis, an alternative matched analysis would have been of interest.

Dr Fraser and his mentors at Johns Hopkins are to be congratulated for undertaking this analysis and for the clarity of the presentation. The message is concise, and primarily limited by the lack of granularity in the STS database. The study leaves us with questions regarding patients who have both a congenital heart defect and a CDH: What is the best timing for repair of the cardiac defect vs the CDH? Are there characteristics of the CDH which make palliation vs complete repair preferable for the heart defect? Are there combinations of heart defect and CDH severity which make intervention futile? In the future, it may be possible

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to link the STS database to the large, multicenter CDH study group database[2] to shed more light on this topic.

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