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Preface: current status of diagnosis and treatment of congenital diaphragmatic hernia

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I am so pleased to present to our readership, this topic collection of the *World Journal of Pediatric Surgery* (*WJPS*) dedicated to congenital diaphragmatic hernia (CDH). It is difficult to think of another congenital anomaly that has inspired more creative collaboration and innovation than CDH. Over the course of 50 years, CDH has gone from being a simplistically understood, postnatally diagnosed surgical emergency with <50% survival,[1] to an antenatally predicted derangement of cardiopulmonary physiology that is selectively amenable to fetal correction and receives expert, team-based, standardized postnatal treatment, with survival rates that approach 90%.[2] The development of CDH-specific international databases like that of the CDH Study Group[3] has accelerated knowledge synthesis, and the growing evidence base has supported the development of standardized practice guidelines spanning the continuum from prenatal diagnosis to long-term follow-up.[4 5]

This compendium of review articles begins with an excellent review of the current state of prenatal diagnosis and postnatal disease severity prediction by Drs Abbasi, Backley, Ryan and Johnson. Accurate fetal CDH risk assessment is essential for parental counseling and treatment planning, including the offer of fetal intervention for those fetuses of highest risk based on prenatal estimation of pulmonary hypoplasia severity. In a companion article by Drs Abbasi, Van Mieghem and Ryan, entitled 'Fetal therapy for CDH: Past, present and future', the history of fetal surgery is summarized from Michael Harrison's proof of concept studies of tracheal occlusion in sheep to the modern era of human fetoscopic endoluminal tracheal occlusion (FETO) following the recently completed randomized controlled trials for severe and moderate-risk fetal CDH. The article concludes with a look forward to minimally invasive fetal therapies that are on the horizon, including transplacental and cell-based therapies.

Drs Liu and Yu have nicely summarized the increased awareness of associated genetic malformations which are observed in 30% of human CDH cases and review the functional investigation of signaling pathways on diaphragm and fetal lung development with gene knockout mouse models. This article also explores the relationship between putative environmental factors including vitamin A and the retinoic acid pathway and its hypothesized regulatory role in gene expression which likely contributes to the developmental CDH phenotype.

Dr Traynor provides an eloquent summary of the evolutionary history of lung-protective ventilation in CDH and uses the significant body of evidence in adult acute respiratory distress syndrome (ARDS) as a platform for understanding respiratory mechanics in hypoplastic lung ventilation and the importance of iatrogenic lung injury avoidance in CDH. The article summarizes the current state of knowledge on the use of conventional and high-frequency modalities and describes the author's own approach to ventilatory management of CDH from birth through to recovery from surgery.

The next article by Drs Surak, Mahgoub and Ting reviews the hemodynamic management of CDH, and highlights the evolving role of targeted neonatal echocardiography performed by specially trained neonatologists in improving bedside recognition of the various phenotypes of cardiopulmonary dysfunction in CDH. The article also provides a comprehensive and contemporary review of the classes of medication used to treat pulmonary hypertension and ventricular dysfunction including pulmonary vasodilators, systemic vasoconstrictors, inotropic agents and prostaglandins.

Despite a necessary emphasis on ventilatory and hemodynamic management of CDH, the role of surgery in determining short- and long-term outcomes is indisputable. Drs Jank,

Boettcher and Keijzer provide a superb summary of the current state of surgical repair for both small and large defects, including a thorough review of the prosthetic and autologous tissue options for diaphragmatic defects not amenable to primary repair. There is also a very nice section on the technical aspects and anesthetic safety considerations for minimally invasive repair.

Drs Gehle, Meyer and Jancelewicz tackle the still controversial subject of the role of extracorporeal life support (ECLS) in severe CDH and review current literature and consensus guidelines in an attempt to address, once and for all, the question of optimal timing of surgery in the infant requiring ECLS support. The thoughtful discussion of this subject highlights the importance of using a strategy that avoids non-repair, which still contributes significantly to mortality in CDH.

With improved survival, the focus of outcome improvement in CDH has appropriately shifted to the health and quality of life outcomes of survivorship, which is only possible if CDH survivors are followed longitudinally by a multidisciplinary team. Drs Cimbak and Buchmiller provide an excellent, state-of-the-art summary of long-term functional surveillance by organ system, including the importance of transitioning to adult care, from the institution that initiated this practice over 30 years ago.

Last, but certainly not least, Drs Dimmer, Baird and Puligandla present a compelling case for the importance of standardization as an essential strategy for achieving optimal outcomes in CDH. The article reviews the multidisciplinary, collaborative and structure-based processes used by the CDH Euro consortium and the Canadian CDH Collaborative in creating their respective, evidence-based consensus guidelines, recommends implementation strategies once guidelines have been developed, and highlights the importance of a commitment to updating guidelines as new evidence emerges.

The progress made in CDH care and outcomes reflects the collaborative innovation of multidisciplinary care teams and scientists, supported by high-quality comparative effectiveness research and quality improvement enabled in part by CDH patient registries. In the years ahead, we can anticipate continued improvements through discovery science and the contributions of artificial intelligence in bringing personalized patient customization to standardized care. On behalf of the editors and editorial staff at *WJPS*, we hope you enjoy this topic collection on CDH.

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REFERENCES

1 Simson JN, Eckstein HB. Congenital diaphragmatic hernia: a 20 year experience. *Br J Surg* 1985;72:733–6.
2 Yang MJ, Fenton S, Russell K, *et al*. Left-sided congenital diaphragmatic hernia: can we improve survival while decreasing ECMO? *J Perinatol* 2020;40:935–42.
3 Holden KI, Ebanks AH, Lally KP, *et al*. The CDH Study Group: Past, Present, and Future. *Eur J Pediatr Surg* 2024;34:162–71.
4 Snoek KG, Reiss IKM, Greenough A, *et al*. Standardized Postnatal Management of Infants with Congenital Diaphragmatic Hernia in Europe: The CDH EURO Consortium Consensus - 2015 Update. *Neonatology* 2016;110:66–74.
5 Puligandla P, Skarsgard E, Baird R, *et al*. Diagnosis and management of congenital diaphragmatic hernia: a 2023 update from the Canadian Congenital Diaphragmatic Hernia Collaborative. *Arch Dis Child Fetal Neonatal Ed* 2024;109:239–52.