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Surgical Management and Outcomes of Congenital Diaphragmatic Hernia in Neonates: A Systematic Review

Dr. Samina Ruquaya[1], Dr. Praveena Gaddam[2], Dr. Athira Gopinathan[*3]

[1]Post graduate Resident, Department of General Surgery, SRM Medical College Hospital and Research Centre, Kattankulathur, Chengalpattu

ORCID ID - 0009-0005-1535-0454

Email ID: saminaruquaya@gmail.com

[2]Post Graduate Resident, Department of General Surgery, SRM Medical College Hospital and Research Centre, Kattankulathur, Chengalpattu

Email ID: g.praveena418@gmail.com

*3Professor, Department of General Surgery, SRM Medical College Hospital and Research Centre, Kattankulathur, Chengalpattu

***Corresponding Author:**

Dr. Athira Gopinathan

Email ID: athirag@srmist.edu.in

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ABSTRACT

Congenital diaphragmatic hernia (CDH) is a severe neonatal disorder characterized by inadequate diaphragm development, leading to pulmonary hypoplasia and hypertension. Notwithstanding advancements in neonatal treatment, congenital diaphragmatic hernia (CDH) is linked to considerable morbidity and mortality. This systematic review assesses contemporary surgical techniques—spanning open and thoracoscopic repair to ECMO-assisted procedures—and their results in newborns with congenital diaphragmatic hernia (CDH). A comprehensive literature review was performed across various databases for studies published from 2000 to 2025. The research indicates that postponing surgical repair following preoperative stabilization results in superior outcomes compared to immediate surgery. Although minimally invasive surgery demonstrates potential for certain stable neonates, open repair continues to be the standard for critically ill patients. Patch repair is frequently essential for substantial flaws but entails increased chances of recurrence. The utilization of ECMO, while life-saving in critical instances, entails long-term neurodevelopmental hazards. Postoperative problems, including pulmonary hypertension, recurrence, and long-term morbidities such as chronic lung disease and developmental disabilities, persist. The prognosis is affected by elements including prenatal severity indicators, liver herniation, genetic abnormalities, and available institutional resources. The review highlights future directions, such as advancements in fetal surgery like FETO, the creation of novel surgical procedures, and the establishment of multidisciplinary follow-up programs to improve the quality of life for survivors. A customized, evidence-driven strategy is crucial for enhancing outcomes in infants with congenital diaphragmatic hernia (CDH).

Keywords: *Congenital diaphragmatic hernia, CDH, neonates, surgical intervention, thoracoscopic repair, ECMO, outcomes, fetal surgery, long-term morbidity, multidisciplinary care*

1. INTRODUCTION

Congenital diaphragmatic hernia (CDH) is an uncommon but life-threatening developmental anomaly marked by the inadequate creation of the diaphragm, permitting abdominal organs to protrude into the thoracic cavity. This structural aberration impairs normal lung development, resulting in different degrees of pulmonary hypoplasia and pulmonary hypertension, both of which significantly contribute to infant morbidity and mortality (Aly et al., 2010). The prevalence of congenital diaphragmatic hernia (CDH) is roughly 1 in 2,500 to 1 in 4,000 live births worldwide (Keijzer & Puri, 2010), and despite progress in neonatal care, the issue remains a significant clinical concern.

The surgical correction of congenital diaphragmatic hernia (CDH) is the fundamental treatment; nevertheless, the scheduling, methodology, and perioperative management measures differ considerably across institutions. Conventional surgical procedures encompass open laparotomy or thoracotomy, but less invasive methods like thoracoscopic repair are becoming increasingly investigated for its prospective advantages regarding recuperation and diminished problems (Okawada et al., 2021). The incorporation of extracorporeal membrane oxygenation (ECMO) in critically ill neonates has enhanced survival rates in specific instances, however it is associated with a heightened risk of long-term problems (Snoek et al., 2016).

Notwithstanding significant clinical progress, mortality rates for CDH may still surpass 30%, especially in instances with concomitant abnormalities or severe pulmonary underdevelopment (Numanoglu et al., 1998). Moreover, survivors may encounter enduring problems, such as chronic pulmonary illness, gastric reflux, cognitive impairments, and growth retardation. These results highlight the significance of a multidisciplinary strategy that encompasses not just surgery but also prenatal diagnosis, enhanced postnatal stabilization, and organized long-term follow-up care.

This systematic review seeks to assess the diverse surgical approaches utilized in the treatment of congenital diaphragmatic hernia (CDH) in newborns, analyze both short- and long-term results, and pinpoint deficiencies in existing therapeutic practices. By conducting a critical study of existing literature, we want to furnish clinicians and researchers with a contemporary comprehension of successful surgical procedures and prognostic determinants affecting infant outcomes in congenital diaphragmatic hernia (CDH).

2. METHODOLOGY

A thorough literature review was performed to find pertinent papers examining the surgical therapy and outcomes of congenital diaphragmatic hernia (CDH) in newborns. Electronic databases such as PubMed, Scopus, Web of Science, and Cochrane Library were systematically queried for research published from January 2000 to March 2025. The subsequent keywords and Medical Subject Headings (MeSH) terms were utilized in diverse combinations: “congenital diaphragmatic hernia,” “CDH,” “neonates,” “surgical management,” “outcomes,” “ECMO,” “thoracoscopic repair,” and “survival rate.” Boolean operators (AND/OR) were employed to enhance the search strategy. Manual examination of the reference lists of qualifying papers was conducted to discover further pertinent studies.

The studies were selected based on the following criteria to ensure relevance and quality:

Eligibility Criteria:

- Research about neonates (0–28 days old) diagnosed with congenital diaphragmatic hernia.
- Research that documented surgical procedures (e.g., open repair, thoracoscopic surgery, ECMO-assisted surgery).
- Articles that presented outcome data, encompassing survival rates, complications, or long-term follow-up.
- Original research publications encompassing randomized controlled trials, cohort studies, case-control studies, and extensive case series involving more than ten patients.
- English publications.

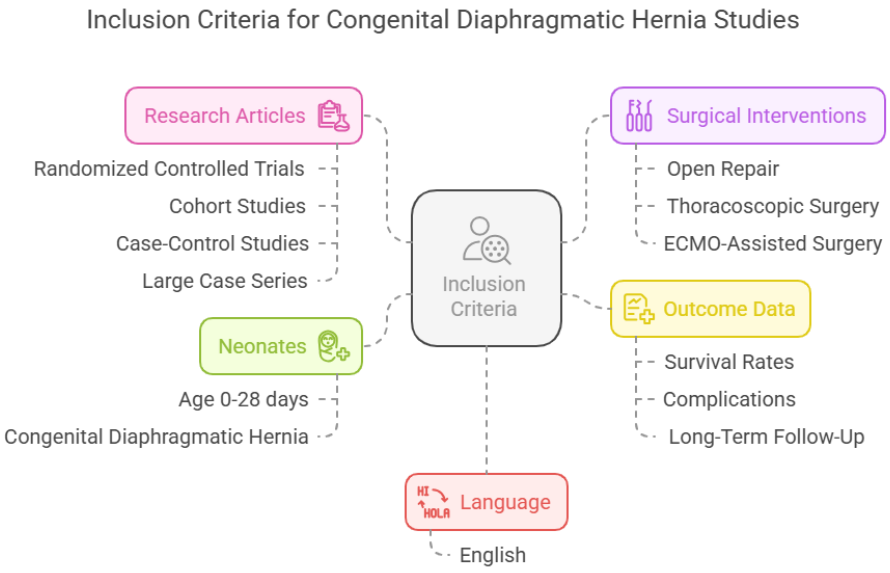

Figure: Inclusion Criteria for Research on Congenital Diaphragmatic Hernia in Neonates

Criteria for Exclusion:

- Research concentrating solely on non-surgical management or fetal interventions.
- Reviews, editorials, letters, or case reports involving less than 10 patients.
- Research lacking definitive outcome metrics or adequate data.
- Articles lacking full-text access or not subjected to peer review.

Data Extraction and Quality Evaluation

Data were separately extracted by two reviewers utilizing a pre-established data extraction sheet. The variables extracted comprised:

- Study attributes (authors, year, country, methodology)
- Sample size and demographic characteristics
- Classification and scheduling of surgical intervention
- Utilization of ECMO or alternative supportive measures
- Documented results: survival rates, surgical complications, long-term morbidity

Standardized techniques were employed to evaluate the quality and risk of bias of the included studies.

- The Newcastle-Ottawa Scale (NOS) for cohort and case-control research
- The Cochrane Risk of Bias Instrument for randomized trials

Discrepancies among reviewers were reconciled by conversation or by consulting a third reviewer. Only papers that exceeded a certain quality criterion were incorporated into the final synthesis to guarantee the robustness and trustworthiness of the findings.

3. SURGICAL INTERVENTION STRATEGIES

The timing of surgical intervention in newborns with congenital diaphragmatic hernia is a crucial determinant that profoundly influences outcomes. Historically, immediate postnatal surgery was conducted; however, this method has predominantly been supplanted by a strategy of delayed surgical intervention, permitting preoperative stabilization of cardiorespiratory function. Contemporary best practices underscore the importance of optimizing oxygenation, ventilation, and perfusion before surgery, especially in instances exacerbated by pulmonary hypertension (Desai et al., 2015). Surgery is generally postponed for 24 to 72 hours, or longer, until the infant achieves hemodynamic stability. This method has been linked to enhanced survival rates and diminished perioperative problems.

Surgical Methodologies (Open versus Minimally Invasive)

Two primary surgical techniques are employed for congenital diaphragmatic hernia (CDH) repair: open surgery (by laparotomy or thoracotomy) and minimally invasive surgery (MIS), chiefly thoracoscopic correction.

- Open surgery continues to be the conventional treatment at numerous facilities, particularly for extremely unwell infants or those need ECMO. It offers enhanced access and visibility, and is seen more dependable for substantial or intricate faults.
- Minimally invasive surgery has become more favored for specific, stable situations owing to its benefits, including diminished postoperative pain, expedited recovery, and reduced infection risk (Nguyen & Le, 2006). Nonetheless, apprehensions persist over elevated recurrence rates and the technical difficulties associated with minimally invasive surgery in small infants.

  The selection of procedure is contingent upon various aspects, such as patient stability, defect dimensions, surgeon proficiency, and institutional guidelines.

Application of Patch Repair and Meshes

The technique of diaphragm closure is a significant surgical aspect.

- Primary closure is optimal and recommended when the diaphragmatic defect is minor and the native tissue can be approximated without strain.
- In instances involving big or "agenetic" diaphragms, patch repair utilizing synthetic or biological meshes is essential. Prevalent materials comprise Gore-Tex and biological patches such as acellular dermal matrices.

Patch repairs effectively restore diaphragm integrity; however, they carry increased risks of recurrence and long-term consequences, including re-herniation and patch infection (Hollinger et al., 2017). Investigations persist in examining more resilient and biocompatible materials for neonatal applications.

Extracorporeal Membrane Oxygenation (ECMO) Assistance

ECMO serves as a life-saving intervention for neonates with congenital diaphragmatic hernia (CDH) experiencing refractory respiratory failure or severe pulmonary hypertension unresponsive to standard mechanical ventilation. ECMO offers transient cardiopulmonary assistance, facilitating stabilization of the lungs and circulatory system prior to surgical intervention.

Nonetheless, ECMO has hazards such as hemorrhage, neurological damage, and infection, necessitating stringent criteria and specialist multidisciplinary teams for its application. Surgery may occur either during ECMO or post-decannulation, contingent upon institutional preference and patient stability. While ECMO has been linked to enhanced survival in critical instances, long-term neurodevelopmental effects continue to be a concern.

4. RESULTS OF SURGICAL INTERVENTION

The surgical correction of congenital diaphragmatic hernia (CDH) is a pivotal event in the treatment of impacted infants; however, outcomes differ significantly based on various criteria, including the degree of pulmonary hypoplasia, concomitant abnormalities, surgical scheduling, and institutional proficiency. This section examines the principal results observed after surgical intervention.

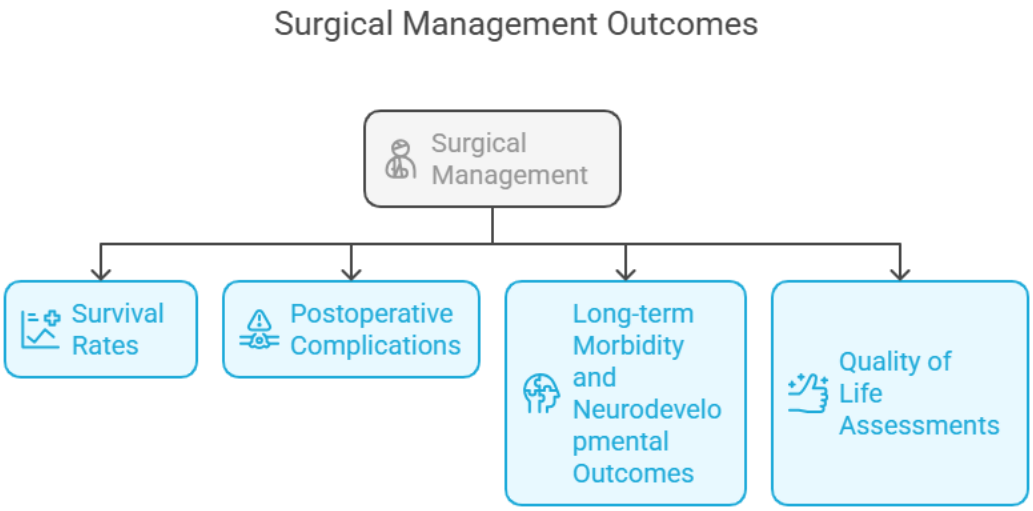

Figure: Essential Elements of Surgical Management Outcomes in Healthcare

Survival Rates

Survival rates in newborns having surgical repair for congenital diaphragmatic hernia (CDH) have markedly improved over recent decades due to breakthroughs in neonatal intensive care, early diagnosis, and surgical methodologies. Recent literature indicates survival rates between 60% and 85% in affluent contexts (Gupta et al., 2023). Survival is significantly affected by disease severity, the occurrence of liver herniation, and the requirement for extracorporeal membrane oxygenation (ECMO). ECMO-supported cases exhibit reduced survival rates, frequently below 50%, however can remain life-saving in extreme situations.

Postoperative Complications

Postoperative problems frequently occur and may encompass pulmonary hypertension, infections, surgical site dehiscence, chylothorax, intestinal obstruction, and hernia recurrence. Recurrence rates range from 10% to 15%, particularly in individuals necessitating patch surgery or possessing substantial diaphragmatic abnormalities (Mattke et al., 2025). The likelihood of problems is elevated in neonates receiving thoracoscopic repair relative to open surgery; yet, the minimally invasive technique has been linked to expedited recovery and reduced hospital durations in some instances.

Chronic Morbidity and Neurodevelopmental Results

Individuals who have undergone CDH surgery frequently experience enduring morbidity, encompassing chronic pulmonary disease, gastroesophageal reflux disease (GERD), inadequate growth, and challenges with feeding (Danzer & Hedrick, 2011). Moreover, neurodevelopmental deficits have been documented in 30%–50% of survivors by the age of two, especially among those who had extended mechanical breathing or ECMO assistance. Cognitive, motor, and linguistic development must be meticulously observed, and early intervention services are frequently essential.

Evaluations of Quality of Life

Although numerous CDH survivors attain satisfactory physical recovery, their quality of life (QoL) may be compromised by ongoing health issues. Recent research employing validated quality of life questionnaires indicate that school-aged survivors of congenital diaphragmatic hernia frequently report restrictions in physical activity, respiratory health, and mental well-being. Parental stress and familial load are critical factors, underscoring the necessity for comprehensive, multidisciplinary long-term follow-up that incorporates psychological and developmental assistance.

5. DETERMINANTS AFFECTING RESULT

The prognosis for infants with congenital diaphragmatic hernia (CDH) is markedly diverse and affected by a confluence of prenatal and postnatal variables. Comprehending these characteristics is essential for risk categorization, parental guidance, and personalized care planning.

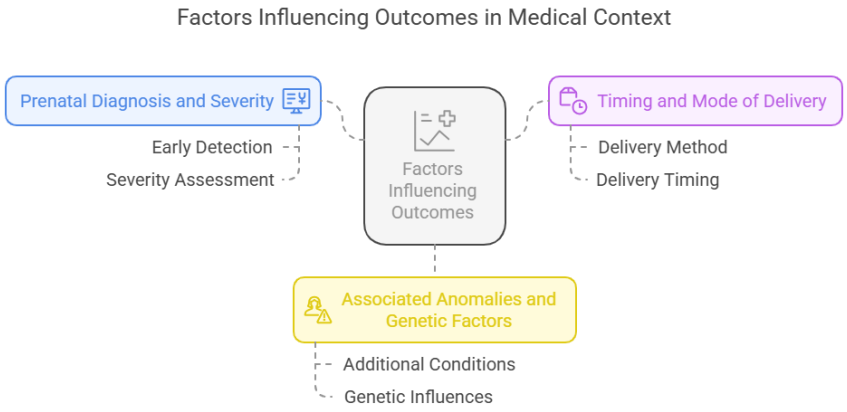

Figure: Principal Determinants of Medical Outcomes: A Framework for Prenatal and Delivery Planning

Prenatal Diagnosis and Severity Assessment

Prenatal diagnosis is crucial in assessing the severity and possible effects of CDH. Most instances are identified during standard fetal abnormality ultrasounds, typically conducted between 18 and 24 weeks of gestation. The lung-to-head ratio (LHR) and its observed-to-expected value (O/E LHR) are critical prognostic indicators that correspond with the degree of lung hypoplasia. An O/E LHR <25% is generally linked to unfavorable postnatal outcomes (Kamata et al., 1992).

Liver herniation into the thoracic cavity, sometimes referred to as "liver-up" cases, signifies a more severe variant of congenital diaphragmatic hernia (CDH) and is associated with increased mortality and morbidity (Albanese et al., 1998). The existence of polyhydramnios, mediastinal displacement, and diminished pulmonary vasculature on prenatal imaging indicates a poorer prognosis.

Timing and Method of Delivery

The ideal timing and method of delivery continue to be topics of discussion, however these elements can affect infant outcomes in congenital diaphragmatic hernia (CDH). Numerous studies indicate that scheduled delivery at a tertiary facility equipped with neonatal intensive care and ECMO capabilities is essential, irrespective of the specific timing (Deprest et al., 2009).

The method of delivery—vaginal or cesarean—has not been conclusively demonstrated to affect survival rates. Nonetheless, cesarean delivery may be deemed appropriate in specific instances to enhance logistics for early postnatal stabilization, especially in fetuses with severe congenital diaphragmatic hernia (CDH). Early elective birth is typically discouraged unless medically warranted, as it may jeopardize fetal lung maturation.

Related Anomalies and Genetic Influences

Approximately 40–60% of congenital diaphragmatic hernia (CDH) instances are linked to concomitant abnormalities, which considerably exacerbate outcomes. The prevalent concomitant problems encompass heart malformations, neural tube defects, and genitourinary anomalies (Veenma, de Klein, & Tibboel, 2012). Complex congenital cardiac disease is a well-established indicator of poor survival.

The genetic factors contributing to CDH have gained increased acknowledgment. Chromosomal anomalies, including trisomy 18, 13, and 21, along with microdeletions (e.g., 15q26), have been associated. Whole exome sequencing has discovered mutations in genes associated with diaphragm development and pulmonary vasculature, such as GATA4, ZFPM2, and FOG2 (Weller et al., 2024). The recognition of syndromic variants of CDH typically correlates with a poorer outcome and informs the severity of postnatal care options.

6. COMPARATIVE ANALYSES AND META-ANALYSES

Comparative studies and multicenter analyses have demonstrated significant regional and institutional disparities in the care and outcomes of congenital diaphragmatic hernia (CDH) in neonates. Divergences arise from resource availability, surgical proficiency, timing of intervention, utilization of ECMO, and newborn intensive care guidelines. High-volume tertiary centers with established multidisciplinary CDH teams frequently demonstrate superior survival rates and reduced postoperative complication rates in comparison to smaller institutions (Martino, Lista, & Guner, 2022). The selection between open repair and minimally invasive techniques varies by geography; thoracoscopic repair is more commonly utilized in centers throughout Europe and North America, whereas certain developing regions predominantly depend on open surgical methods due to restricted access to advanced laparoscopic technology and expertise.

Socioeconomic and healthcare infrastructural elements also affect post-operative results. In low- and middle-income countries (LMICs), delayed diagnosis, inadequate newborn transport facilities, and absence of ECMO result in markedly elevated mortality rates. A comparative analysis of outcomes across continents revealed that infant mortality due to congenital diaphragmatic hernia (CDH) exceeded 40% in low- and middle-income countries (LMICs), in contrast to 15–20% in rich nations (Lum et al., 2022). These differences highlight the necessity for worldwide standardization of treatment methods and capacity-building initiatives in resource-constrained environments.

Temporal Trends in Surgical Outcomes

In the last twenty years, numerous meta-analyses and longitudinal studies have evidenced an improvement in surgical outcomes for CDH, largely attributable to breakthroughs in neonatal care, enhanced ventilatory tactics, earlier prenatal diagnosis, and more sophisticated surgical techniques. Mortality rates, previously surpassing 50%, have decreased in numerous places to around 20% for isolated cases of congenital diaphragmatic hernia without significant abnormalities (Thodika et al., 2022).

The implementation of postponed surgery—facilitating preoperative stabilization of pulmonary function—has been a significant turning point. Previous treatments prioritized prompt surgical intervention; however, recent evidence indicates that delaying surgery for 24–48 hours to optimize cardiopulmonary function results in improved postoperative outcomes (Cox et al., 2022). Furthermore, advancements like as gentle ventilation, permissive hypercapnia, and lung-protective methods have significantly enhanced perioperative treatment.

Meta-analyses have shown a decrease in long-term morbidities, including chronic lung disease and neurodevelopmental impairments, particularly in newborns treated at specialist clinics with organized follow-up programs. Consequently, survival is no longer the exclusive emphasis of CDH management; quality of life and long-term developmental outcomes are increasingly recognized as essential indicators of success.

7. PROSPECTIVE AVENUES AND RESEARCH REQUIREMENTS

Progress in Fetal Surgical Techniques

Recent years have seen substantial advancements in fetal surgical procedures for severe congenital diaphragmatic hernia, especially in fetuses exhibiting high-risk prognostic indications. Fetoscopic Endoluminal Tracheal Occlusion (FETO) is a highly promising technique that entails the temporary obstruction of the embryonic trachea to promote lung development by retaining lung fluid. Randomized controlled trials, such as the TOTAL study (Deprest et al., 2021), have shown that FETO may enhance survival in fetuses with severe pulmonary hypoplasia, but it is a complex and specialized technique. Continued research is essential to enhance timing, improve patient selection criteria, and mitigate risks such as preterm labor and membrane rupture. As fetal surgery advances, coordinated worldwide registries and long-term outcome data will be crucial for establishing uniform protocols and enhancing accessibility to this life-saving procedure.

Advancements in Surgical Methodologies

The surgical care of congenital diaphragmatic hernia (CDH) has historically depended on open repair; however, the growing

utilization of less invasive procedures, including thoracoscopic and laparoscopic methods, signifies a notable transformation in clinical practice. These procedures provide advantages including less surgical discomfort, abbreviated hospitalizations, and enhanced aesthetic results. Nonetheless, apprehensions remain about recurrence rates and the significant learning curve linked to these techniques (Okawada et al., 2021). Subsequent research ought to concentrate on comparison studies to assess the efficacy and safety of these approaches across diverse patient populations and institutional contexts. Furthermore, innovations in biomaterials for patch repair, robotic-assisted surgery, and customized surgical planning with 3D modeling are promising domains that merit additional investigation.

Multidisciplinary Monitoring Initiatives

Individuals who survive CDH frequently have enduring problems, including as chronic pulmonary disease, cognitive impairments, gastrointestinal issues, and growth anomalies. This underscores the necessity for organized, multidisciplinary follow-up programs that incorporate neonatologists, pulmonologists, neurologists, dietitians, and developmental therapists. Such programs should emphasize not just physical rehabilitation but also cognitive, emotional, and social advancement. Recent evidence underscores the need of standardized follow-up protocols in the early detection and intervention of developmental impairments (Pollack et al., 2024). Future projects should prioritize the establishment of extensive national or international CDH follow-up registries to inform best practices and enhance the quality of life for survivors and their families.

8. CONCLUSION

Congenital diaphragmatic hernia in neonates presents intricate therapeutic problems, necessitating prompt diagnosis, tailored surgical approaches, and thorough postoperative management. This review emphasizes the significance of postponing surgical intervention until hemodynamic stabilization is achieved, the selective implementation of minimally invasive procedures, and the prudent use of ECMO in severely unwell newborns. Despite advancements in survival rates, long-term consequences include pulmonary morbidity, neurodevelopmental abnormalities, and recurrence continue to pose substantial concerns. A multidisciplinary, patient-centered strategy—incorporating improvements in fetal surgery, surgical innovation, and systematic long-term follow-up—is crucial for improving both survival rates and quality of life. Ongoing research, standardized treatment protocols, and international collaboration will be crucial in bridging existing gaps and enhancing care for infants with CDH.

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