DIAGNOSIS AND MANAGEMENT OF NEONATAL ESOPHAGEAL ATRESIA: A SYSTEMATIC REVIEW OF CURRENT EVIDENCE

DIAGNÓSTICO E MANEJO DA ATRESIA ESOFÁGICA NO PERÍODO NEONATAL: UMA REVISÃO SISTEMÁTICA DAS EVIDÊNCIAS ATUAIS

DIAGNÓSTICO Y MANEJO DE LA ATRESIA ESOFÁGICA EN EL PERÍODO NEONATAL: UNA REVISIÓN SISTEMÁTICA DE LA EVIDENCIA ACTUAL

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ABSTRACT

Neonatal esophageal atresia is characterized by diagnostic complexity and the need for specialized clinical and surgical management, playing an important role in reducing neonatal morbidity and mortality. This study aims to analyze the available evidence regarding the diagnosis and management of esophageal atresia in newborns, emphasizing early identification, timely intervention, and multidisciplinary care. This is a systematic review conducted according to PRISMA recommendations and registered in PROSPERO under number CRD420261361849, with searches performed in the PubMed, LILACS/BVS, and SciELO databases from 2016 to 2026. The findings demonstrated that early diagnosis, especially during the prenatal period, associated with postnatal clinical and radiological confirmation, reduces complications. Furthermore, early surgical management combined with specialized neonatal intensive care contributes to better clinical outcomes and increased survival. It is concluded that the standardization of diagnostic and therapeutic approaches, together with strengthened multidisciplinary care, is essential to improve neonatal prognosis.

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Keywords: Esophageal Atresia. Newborn. Diagnosis. Treatment. Neonatal Care.

RESUMO

A atresia de esôfago no período neonatal destaca-se pela complexidade diagnóstica e pelo manejo clínico-cirúrgico especializado, sendo relevante para a redução da morbimortalidade neonatal. Objetiva-se analisar as evidências acerca do diagnóstico e manejo da atresia de esôfago em recém-nascidos, enfatizando a identificação precoce, a intervenção oportuna e a assistência multiprofissional. Trata-se de uma revisão sistemática conduzida conforme as recomendações do PRISMA e registrada no PROSPERO sob o número CRD420261361849, com buscas nas bases PubMed, LILACS/BVS e SciELO, no período de 2016 a 2026. Observou-se que o diagnóstico precoce, especialmente no pré-natal, associado à confirmação clínica e radiológica pós-natal, reduz complicações. Além disso, o manejo cirúrgico precoce aliado aos cuidados intensivos neonatais especializados favorece melhores desfechos clínicos e maior sobrevida. Conclui-se que a padronização das condutas diagnósticas e terapêuticas, associada ao fortalecimento da assistência multiprofissional, é fundamental para melhorar o prognóstico neonatal.

Palavras-chave: Atresia Esofágica. Recém-Nascido. Diagnóstico. Tratamento. Cuidados Neonatais.

RESUMEN

La atresia esofágica en el período neonatal se destaca por la complejidad diagnóstica y el manejo clínico-quirúrgico especializado, siendo relevante para la reducción de la morbimortalidad neonatal. Este estudio tiene como objetivo analizar la evidencia disponible sobre el diagnóstico y manejo de la atresia esofágica en recién nacidos, enfatizando la identificación precoz, la intervención oportuna y la atención multiprofesional. Se trata de una revisión sistemática realizada conforme a las recomendaciones PRISMA y registrada en PROSPERO bajo el número CRD420261361849, con búsquedas en las bases de datos PubMed, LILACS/BVS y SciELO, en el período de 2016 a 2026. Los hallazgos demostraron que el diagnóstico precoz, especialmente en el período prenatal, asociado a la confirmación clínica y radiológica posnatal, reduce las complicaciones. Además, el manejo quirúrgico temprano combinado con cuidados intensivos neonatales especializados favorece mejores resultados clínicos y mayor supervivencia. Se concluye que la estandarización de las conductas diagnósticas y terapéuticas, asociada al fortalecimiento de la atención multiprofesional, es fundamental para mejorar el pronóstico neonatal.

Palabras clave: Atresia Esofágica. Recién Nacido. Diagnóstico. Tratamiento. Cuidados Neonatales.

1 INTRODUCTION

Esophageal atresia (EA) is one of the most frequent congenital malformations of the gastrointestinal tract, with an estimated incidence of between 1 in 2,500 and 4,500 live births. It is characterized by the interruption of the continuity of the esophagus, often associated with tracheoesophageal fistula, and may occur in isolation or as part of complex syndromes1. It is a condition of high clinical complexity, whose management requires early and articulated interventions, given its association with significant morbidity and mortality in the neonatal period.

Early diagnosis is a central element for the proper management of cases, allowing the planning of delivery in referral centers with intensive neonatal support and the early organization of care. Evidence indicates that delays in EC recognition and surgical intervention are associated with a higher risk of complications and mortality, reinforcing the importance of care protocols that favor the timely identification and referral of newborns2,3.

Despite advances in perioperative care, EC still has a high complication rate. Clinical studies have shown the occurrence of adverse events, such as anastomotic stenosis, anastomotic rupture, and recurrent fistula, even in specialized services, evidencing the need for rigorous monitoring and well-established clinical strategies4. In this context, factors such as prematurity, the presence of associated malformations, and the clinical conditions of the newborn directly influence the outcomes.

The management of esophageal atresia in the neonatal period involves the integration of intensive care, surgical intervention, and ongoing clinical support. Strategies such as ventilatory support, secretion control, progressive enteral nutrition, and neonatal unit monitoring have been associated with better clinical outcomes [5,6,7]. In addition, the proper management of the perioperative period is a determining factor for recovery and for the prevention of short- and long-term complications.

In more complex situations, such as cases without fistula or associated with other malformations, multidisciplinary action is essential. The articulation between neonatology, pediatric surgery, cardiology, nutrition, and other health fields contributes to clinical stabilization, management of comorbidities, and continuity of care. This integrated approach is also related to the organization of care networks, which are considered fundamental to reduce inequalities in access to specialized care and improve clinical outcomes8.

In this context, there is a need to strengthen strategies that promote early diagnosis, adequate clinical-surgical management, and integration between different levels of care. This study analyzes the diagnosis and management of esophageal atresia in the neonatal period, with emphasis on early identification, clinical and surgical strategies, and care organization.

In addition, it examines risk factors and perioperative complications, as well as discusses the role of multidisciplinary approaches and care networks in improving neonatal outcomes.

2 METHODS

This is a systematic review of the literature, with a qualitative approach, conducted with the objective of analyzing the diagnosis and management of esophageal atresia in the neonatal period. The study was developed according to the recommendations of PRISMA 20209, ensuring methodological rigor in the stages of identification, selection, and analysis of the studies.

The search was carried out in the PubMed, LILACS/VHL and SciELO databases, using descriptors related to esophageal atresia, neonatal population and diagnostic and therapeutic aspects. In the PubMed database, the following terms were used: ("Esophageal Atresia") AND ("Newborn" OR "Neonate") AND ("Diagnosis" OR "Treatment" OR "Management"). In the LILACS/VHL and SciELO databases, descriptors in Portuguese and Spanish were used: ("Esophageal Atresia") AND ("Newborn" OR "Born Child") AND ("Diagnosis" OR "Treatment" OR "Treatment"), in order to increase the sensitivity of the search.

The identified records were exported to the Rayyan platform, where duplicates were removed using an automated tool, followed by manual verification based on title, authorship, year of publication, and DOI.

Studies addressing esophageal atresia in the neonatal period, focusing on diagnosis and clinical and/or surgical management, published between 2016 and 2026, in Portuguese, English, or Spanish, and with full text available, were included. Original studies, including cohorts, case-control studies, cross-sectional studies, and case series, were considered eligible.

Duplicate studies, literature reviews, editorials, letters to the editor, and event summaries were excluded, as well as those that were not directly related to the topic or that included populations outside the neonatal period. Articles that did not specifically address aspects of diagnosis or management of esophageal atresia or that were not available in full were also excluded.

After selection, the studies were submitted to analytical reading and organized into thematic axes related to early diagnosis, clinical-surgical management, and care organization. The synthesis of the data was carried out qualitatively, with a critical interpretation of the available evidence.

As this is a secondary study, based on data in the public domain, there was no need to submit it to the Research Ethics Committee, in accordance with current regulations.

3 RESULTS

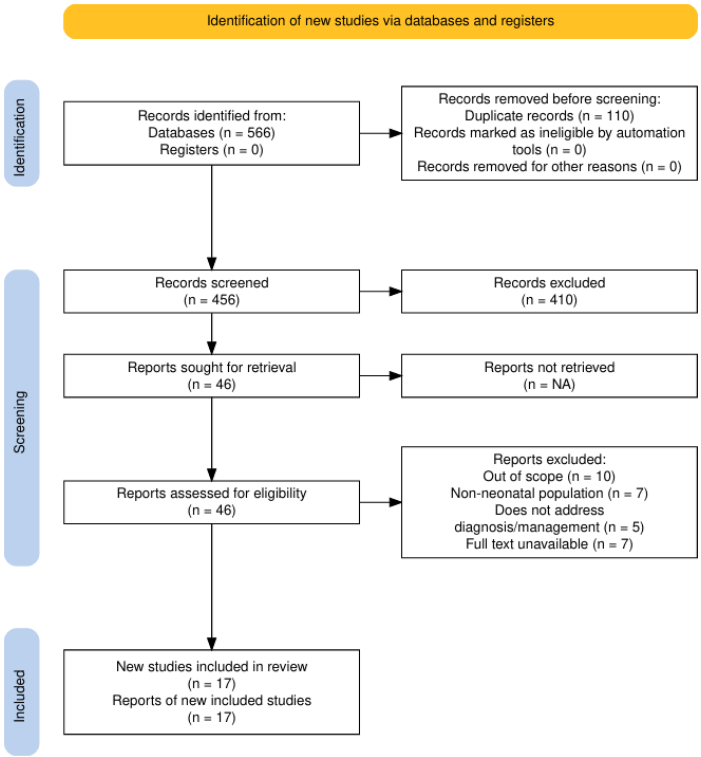
The search in the databases resulted in the identification of 566 records, from PubMed (n=489), LILACS/VHL (n=68) and SciELO (n=9). After the removal of 110 duplicates, 456 studies remained for the screening stage.

The screening, carried out through the reading of titles and abstracts, resulted in the exclusion of 410 records for not meeting the previously established eligibility criteria. Thus, 46 studies were selected for full reading. After the evaluation of the full texts, 29 articles were excluded, mainly because they did not fit the scope of the research, included populations outside the neonatal period, did not address aspects of diagnosis or management of esophageal atresia, or did not present full text availability. At the end of the process, 17 studies were included in the review.

The process of identification, screening, eligibility, and inclusion of studies is presented in Figure 1, prepared according to the recommendations of PRISMA 20209.

Figure 1

Flowchart of the selection process of studies according to PRISMA 2020



Source: Authors.

The included studies were published between 2017 and 2026, with a predominance of observational designs, including retrospective, cohort, and cross-sectional studies. Although the time frame of the search covered the period from 2016 to 2026, no eligible studies were identified in the initial year according to the established criteria. The samples varied considerably, from case series with small numbers of participants to multicenter studies with more than a thousand patients, reflecting the heterogeneity of the scientific production on esophageal atresia in the neonatal period. The detailed characterization of the included studies is described in Table 1.

Table 1

Characterization of included studies

Author/Year	Country	Design	Sample	Objective	Key findings
Piro et al. (2018)	Italy	Retrospective	67 Patients	To evaluate clinical features and their association with morbidity and mortality	Higher mortality associated with congenital malformations and need for intensive care
Tröbs et al. (2017)	Germany	Observational	24 Patients	To compare postoperative course, morbidity, and early outcomes between isolated TEF and esophageal atresia with distal TEF	High rates of prematurity and congenital malformations; Increased perioperative morbidity in esophageal atresia
Basuguy et al. (2020)	Turkey	Retrospective	98 patients	Evaluate the management of fistula and complications (extravasation)	Long esophageal gap associated with higher rates of complications and morbidity
Serooskerken et al. (2021)	Netherlands	Observational	64 patients	Analyze thoracoscopic repair	Surgery considered safe, however, associated with complications such as stenosis and extravasation
Agurto-Ramírez et al. (2023)	Spain	Transverse	146 Patients	Assess prevalence and associated factors	Strong association with congenital malformations and low birth weight
Fola et al. (2023)	Cameroon	Prospective	6 patients	Analyze management strategies in	Survival possible with adapted

				low-resource contexts	techniques, despite structural limitations
Odera et al. (2023)	South Africa	Retrospective	180 patients	Assess incidence and clinical outcomes	Survival rate of 70%; outcomes influenced by birth weight and sepsis
Borselle et al. (2024)	Poland	Retrospective cohort	145 patients	To evaluate the thoracoscopic approach according to birth weight	Technique considered safe even in patients with low birth weight
Kaya et al. (2024)	Turkey	Retrospective	10 patients	To analyze surgical techniques for long-gap esophageal atresia	Techniques such as Foker and Gazi proved to be effective, but associated with complications
Hall et al. (2024)	United Kingdom	Retrospective	157 patients	Evaluate a minimally interventionist approach	Early enteral feeding improves recovery
Pandya et al. (2025)	India	Observational	100 patients	Analyze diagnosis and management	Survival influenced by sepsis, birth weight, and associated anomalies
Ali et al. (2025)	Saudi Arabia	Cohort	67 patients	Assess the impact of esophageal gap	Long esophageal gap associated with increased morbidity and prolonged recovery
Khosravi et al. (2025)	Iran	Retrospective	115 Patients	Compare surgical techniques	Similar outcomes between primary and staged approaches
Platt et al. (2025)	Canada	Retrospective cohort	27 Patients	Evaluate multiprofessional care	Multiprofessional care associated with better care coordination and reduction of hospitalizations
Ungruh et al. (2025)	Germany	Observational	1475 patients	Evaluate the organization of the health system	High complication rates reinforce the importance of centralizing care and specialized centers
Alizadeh et al. (2026)	Iran	Retrospective	79 Patients		Routine chest drainage associated with

				Evaluate the use of chest drainage	increased complications and ICU length
Lindeboom et al. (2026)	Netherlands	Cohort/RCT in development	38 Patients	Evaluate for primary tracheopexy	Reduction of respiratory infections and morbidity

Source: Prepared by the authors

In general, it has been evidenced that esophageal atresia remains a complex condition, often associated with congenital malformations and clinical factors that directly impact the prognosis, such as prematurity, low birth weight, and the presence of comorbidities10,11,12,13,14.

Early diagnosis, usually performed in the first hours or days of life, has been identified as a determining factor for better clinical outcomes, being associated with a reduction in complications and neonatal mortality13,14,10.

With regard to management, all studies indicate surgical correction as the standard treatment, with different technical approaches employed according to the anatomical and clinical characteristics of the patients. In three studies, it was observed that the results tend to be similar between different techniques when performed under appropriate conditions1,15,16. On the other hand, three studies have highlighted that the length of the esophageal gap, the presence of infection, and neonatal clinical instability are determining factors for postoperative complications17,18,19.

Within the scope of perioperative strategies, three studies showed a direct impact of interventions on clinical outcomes. Routine use of chest drainage was associated with increased complications and longer hospital stays20, while early enteral feeding demonstrated benefits in recovery6. In addition, primary tracheopexy has shown promising results in reducing respiratory morbidities in selected patients21.

Regarding the organization of care, four studies indicated that the multidisciplinary approach, the centralization of care, and the structuring of care systems are associated with improved clinical outcomes, including longer survival and reduced length of hospital stay8,14,21,22. On the other hand, one study showed that, in contexts with limited resources, although it is possible to achieve survival, significant structural challenges persist that negatively impact the results22.

The synthesis of the findings allowed the organization of the evidence into three main thematic axes: early diagnosis and clinical evaluation, neonatal clinical-surgical management, and multidisciplinary care networks, as shown in Table 2.

Table 2

Thematic axes of scientific production on neonatal esophageal atresia

Thematic axis	Key Evidence	Summary of findings
Early diagnosis and clinical evaluation (n = 5)	Identification in the first hours of life, characteristic clinical signs and confirmation by imaging tests; high association with congenital malformations	Early diagnosis allows immediate intervention, reduces complications, and improves neonatal prognosis, especially in patients with associated comorbidities
Neonatal clinical and surgical management (n = 8)	Initial stabilization, surgical techniques (primary anastomosis and staged repairs), influence of the esophageal gap, postoperative complications, and perioperative strategies	Individualized management, considering clinical and anatomical conditions, is associated with better outcomes; interventions such as chest drainage should be judicious, while new surgical approaches show potential benefit
Multidisciplinary care networks (n = 4)	Integration between neonatology, pediatric surgery and other specialties; Centralisation in specialised centres	The organization of care and multiprofessional action contribute to the reduction of complications, improved survival and higher quality of care

Source: Prepared by the authors

4 DISCUSSION

This systematic review, by integrating evidence from 17 original studies, confirms that esophageal atresia in the neonatal period remains a condition of high clinical complexity, whose evolution results from the interaction between biological, care, and structural factors. Although advances in diagnosis and management have contributed to the improvement of outcomes, important inequalities persist related to the organization of health services, access to specialized centers, and the standardization of care.

The findings show convergence in the literature regarding the relevance of early diagnosis as a determinant for reducing morbidity and mortality. Studies conducted in different contexts have shown that identification in the first hours of life, associated with the systematic evaluation of congenital malformations and clinical factors, is directly related to better outcomes [10,12,13]. However, it is observed that this diagnostic capacity is not homogeneous among the services, being influenced by the availability of resources, team training and organization of the care network, which evidences an important component of health inequity.

In the context of clinical-surgical management, the results indicate relative consistency among studies regarding the efficacy of different surgical approaches when performed in appropriate contexts1,15,16. However, this apparent equivalence should be interpreted with caution, since factors such as esophageal gap length, prematurity, and the presence of comorbidities exert a significant influence on outcomes17,18. In addition, variability in the indication criteria and in the experience of the teams may contribute to

heterogeneous results, indicating the need for greater standardization and more robust comparative studies.

The analysis of perioperative practices reveals a changing field, in which traditionally consolidated interventions have been questioned in the light of recent evidence. The routine use of thoracic drainage, for example, was associated with a higher incidence of complications and prolonged hospitalization20, suggesting that its application should be judicious and individualized. On the other hand, strategies such as early enteral feeding6 and innovative interventions, such as primary tracheopexy21, point to a movement to improve care practices, although they lack validation in multicenter studies and randomized clinical trials.

The organization of care emerges as one of the main determinants of outcomes, transcending strictly clinical aspects. Consistent evidence indicates that the centralization of care in specialized centers and the performance of multidisciplinary teams are associated with a reduction in complications, longer survival, and better coordination of care8,22. On the other hand, studies conducted in contexts with limited resources show that, although technical adaptations can guarantee survival, relevant structural barriers persist that negatively impact the results23. These findings reinforce that the management of esophageal atresia should be understood not only as a clinical challenge, but also as a matter of organization of health systems.

Despite the advances observed, this review highlights important limitations in the literature, including the predominance of observational studies, methodological heterogeneity, and the lack of standardization of care protocols. These limitations restrict the comparability of results and hinder the consolidation of more robust evidence, especially with regard to the definition of best clinical and organizational practices.

Thus, the findings indicate that the management of esophageal atresia in the neonatal period should be guided by an integrated approach, which articulates early diagnosis, individualized clinical decision-making, and efficient organization of health services. The strengthening of care networks, the centralization of care, and the implementation of evidence-based protocols are fundamental strategies for reducing inequalities and improving neonatal outcomes.

5 CONCLUSION

This systematic review shows that esophageal atresia in the neonatal period remains a highly complex condition, whose management requires integration between early diagnosis, appropriate surgical decision, and efficient organization of care. Acting in the first

hours of life, associated with comprehensive clinical evaluation, is a central element for reducing complications and improving neonatal outcomes.

In the therapeutic field, it is observed that, although different surgical approaches present similar results in appropriate contexts, the individualization of management, considering anatomical characteristics, clinical conditions, and comorbidities, is determinant for the evolution of patients. In addition, perioperative practices have been reevaluated in the light of new evidence, indicating the need for continuous updating of conducts.

The organization of care stands out as a fundamental component, since the multidisciplinary approach and centralization in specialized centers are associated with greater care safety, continuity of care, and better clinical outcomes. In this sense, the structure of health services and access to care networks are determining factors for the quality of neonatal care.

However, limitations in the literature persist, including methodological heterogeneity, lack of standardization of protocols, and scarcity of prospective and multicenter studies, which restricts the comparability of findings and the consolidation of more robust evidence.

It is concluded that the management of esophageal atresia in the neonatal period should be guided by an integrated, individualized and evidence-based approach, considering both the clinical-surgical aspects and the organization of health services. The strengthening of care networks and the production of studies with greater methodological rigor are fundamental for the reduction of care inequalities and the continuous improvement of the quality of neonatal care.

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