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Review Article

Pulmonary Sequelae of Congenital Heart Disease: A Growing Challenge Across Systems

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Received: January 27, 2026; **Published:** February 16, 2026

Abstract

As survival rates for congenital heart disease (CHD) continue to improve, long-term pulmonary comorbidities, such as chronic lung disease (CLD), are emerging as critical challenges across the lifespan. This review explores the intricate cardiopulmonary interplay, beginning in fetal development, in which disrupted hemodynamics impair lung growth and maturation. It further examines how surgical interventions, mechanical ventilation, and environmental factors compound pulmonary injury, leading to a spectrum of sequelae including pulmonary hypertension, airway malacia, and restrictive lung physiology. It is not just patients who struggle; families, too, are often overwhelmed by emotional stress, financial strain, and the exhausting logistics of prolonged hospital stays and complex home care. Emphasizing the need for an integrated, lifespan-oriented model of care, the article advocates for early detection, preventive strategies, mental health support, and global health equity to mitigate the rising burden of CLD in CHD populations.

Keywords: *Pulmonary Sequelae; Congenital Heart Disease (CHD); Chronic Lung Disease (CLD)*

Introduction

Over the past several decades, remarkable advancements in surgical and medical care have transformed the landscape of congenital heart disease (CHD). Children born with complex cardiac anomalies are surviving into adolescence and adulthood at increasing rates [1]. However, improved survival has shifted the clinical focus from short-term postoperative outcomes to long-term comorbidities, with chronic lung disease (CLD) emerging as a particularly complex and often underrecognized consequence of CHD.

Chronic lung disease in the setting of CHD is not a peripheral or secondary issue. Rather, it is an intrinsic extension of congenital cardiac pathology, arising from a combination of altered development, surgical interventions, hemodynamic changes, and environmental exposures. Recognition of this interdependence is critical to optimizing lifelong outcomes for patients with CHD.

Methods: Literature Review Approach

This article is a narrative review intended to synthesize contemporary concepts related to pulmonary sequelae in congenital heart disease rather than to provide a formal systematic review. Relevant literature was identified through a targeted review of peer-reviewed publications using commonly used databases, including PubMed/MEDLINE and Google Scholar.

The literature search was conducted to capture clinically and mechanistically relevant studies addressing cardiopulmonary development, chronic lung disease, pulmonary hypertension, airway abnormalities, functional outcomes, and global health considerations in congenital heart disease. Priority was given to recent publications, consensus statements, and clinical guidelines, with inclusion of earlier seminal studies when necessary to provide historical or developmental context. Articles were selected based on relevance to cardiopulmonary pathophysiology, clinical outcomes, and health systems implications rather than predefined quantitative inclusion or exclusion criteria.

Developmental interdependence: Cardiopulmonary interactions

The embryological development of the heart and lungs is closely connected, governed by shared signaling pathways and spatial coordination during critical periods of organ formation. Both systems originate from mesodermal tissues and develop in parallel, with structural and functional maturation occurring in a time-sensitive and interdependent manner. This relationship ensures that the pulmonary and cardiovascular systems are aligned for efficient gas exchange and circulation after birth. In CHD, this developmental synchrony is often disrupted. Structural abnormalities, particularly complex defects, alter fetal and early neonatal circulation. These changes result in reduced or abnormal pulmonary blood flow during critical phases of lung development, particularly during the canalicular and saccular stages, when alveoli and pulmonary vessels are forming and maturing.

Evidence from both animal models and human autopsy studies has shown that diminished pulmonary perfusion during these stages leads to significant disruption of lung development [2]. The affected lung often shows simplified alveolar structures, reduced capillary density, and disorganized connective tissue, resembling the histological pattern of bronchopulmonary dysplasia. The result is a structurally and functionally immature lung with impaired gas exchange, increased vascular resistance, and limited respiratory reserve [3].

The management of neonates with CHD frequently introduces additional stressors. Many infants with complex heart defects require prolonged mechanical ventilation, high concentrations of supplemental oxygen, and cardiopulmonary bypass in the first days or weeks of life. While these therapies are essential for survival, they introduce risks such as oxygen toxicity, ventilator-associated lung injury, and systemic inflammation. For example, oxygen therapy can impair surfactant production and increase oxidative stress, while positive-pressure ventilation may lead to alveolar overdistension and barotrauma. Cardiopulmonary bypass itself is associated with an inflammatory response that can affect both the alveolar epithelium and pulmonary vasculature [4,5].

These factors contribute to a chronic injury pattern involving pulmonary vascular remodeling, interstitial fibrosis, and increased risk of pulmonary hypertension. Over time, this process may result in a chronic lung disease phenotype that limits respiratory function and physical endurance. The way these two systems develop and fall apart defines everything that follows. Without that understanding, we are always a step behind. In some cases, early surgical repair can help, but only if it restores pulmonary blood flow before the window for lung development closes. Similarly, lung-protective ventilation strategies and careful oxygen management during critical care can help reduce the risk of further developmental injury.

Surgical and structural contributors to pulmonary dysfunction

Surgical palliation and repair are central to the management of complex CHD. However, they also contribute significantly to pulmonary sequelae. In single-ventricle physiology, the Glenn and Fontan procedures reroute systemic venous return directly to the pulmonary arteries, resulting in non-pulsatile, passive pulmonary blood flow. Over time, this abnormal hemodynamic state leads to increased pulmonary vascular resistance, reduced compliance, ventilation-perfusion mismatch, and restrictive lung mechanics [1,6].

Over time, the body pays a price. Multiple surgeries and tubes can leave behind scarring, nerve damage, and a chest wall that no longer moves as it should. These mechanical changes further restrict lung expansion and reduce pulmonary reserve, particularly in patients with prolonged postoperative recovery or repeated interventions [7,8].

It is surprisingly common. Patients with CHD often have airways that collapse or get compressed by nearby structures. Dilated cardiac chambers, vascular grafts, or enlarged pulmonary arteries can exert external pressure on the trachea or mainstem bronchi, producing dynamic airway collapse or fixed obstruction. Additionally, congenital airway anomalies such as tracheobronchomalacia may be unmasked or worsened by repeated intubation and prolonged ventilator dependence [9].

Pulmonary hypertension: A devastating sequela

Among the most severe pulmonary complications in CHD is pulmonary arterial hypertension (PAH). This condition may arise from chronic left-to-right shunting, elevated left atrial pressures, pulmonary venous obstruction, or chronic hypoxemia. Persistent elevation in pulmonary pressures triggers a maladaptive cascade of vascular remodeling, intimal proliferation, and smooth muscle hypertrophy. If left untreated, this can culminate in Eisenmenger syndrome, a life-limiting condition marked by shunt reversal, cyanosis, and right heart failure [10].

While targeted pharmacotherapy (such as endothelin receptor antagonists, phosphodiesterase inhibitors, and prostacyclin analogs) has improved outcomes in idiopathic PAH, their use in CHD-associated PAH requires tailored assessment. Recent guidelines emphasize early recognition, individualized management, and the inclusion of pediatric patients in clinical trials to advance the evidence base [1,11]. Pulmonary vascular resistance and compliance are emerging as valuable prognostic markers in this population. The resistance-capacitance relationship provides insights into vascular stiffness and right ventricular load, two factors that are crucial in long-term risk stratification [12].

Functional and psychosocial impact

For many children with CHD, even simple activities become exhausting; chronic lung disease steadily chips away at their stamina and freedom to move [5,13]. Beyond physical symptoms, the psychosocial burden is substantial and varies across developmental stages.

In infancy, frequent hospitalizations and invasive procedures may lead to feeding and sleeping difficulties, hypersensitivity to stimuli, and early developmental delays, often exacerbated by repeated separations from caregivers. School-aged children commonly experience social withdrawal, anxiety, depression, or behavioral challenges related to missed school and peer interactions, with increased risk for attention difficulties and cognitive impairment. Adolescents face distinct challenges, including struggles with independence, body image concerns, and frustration related to physical limitations as they transition to adult care. In adulthood, survivors may contend with progressive symptoms that affect employment, finances, family planning, and relationships, alongside heightened concerns about mortality. Across the lifespan, individuals with CHD demonstrate an increased prevalence of mood and anxiety disorders, with lifetime rates approaching 50%, exceeding those observed in the general population [14-16].

The psychosocial burden extends to families, who face emotional stress and financial strain related to recurrent hospitalizations, prolonged recovery periods, and complex home care requirements [17,18]. And yet, mental health care often gets sidelined. Rehabilitation is too rarely part of the plan, even when the need is obvious. Pediatric pulmonary rehabilitation programs, incorporating supervised exercise, airway clearance techniques, and respiratory muscle training, have demonstrated benefits in other chronic lung diseases but remain underutilized in CHD populations [4]. Integrating age-appropriate mental health support, including psychotherapy, peer support, and carefully selected pharmacologic interventions, is essential to improving quality of life and long-term outcomes [14].

Disparities in low-resource settings

The burden of chronic lung disease in congenital heart disease is disproportionately high in low- and middle-income countries (LMICs), where delayed diagnosis and limited access to surgical care prolong exposure to abnormal cardiopulmonary hemodynamics. Environmental factors such as air pollution, recurrent respiratory infections, malnutrition, and overcrowded living conditions further compound pulmonary injury and worsen outcomes [6,19].

In LMICs, unoperated or late-treated CHD frequently leads to severe pulmonary complications, including pulmonary hypertension, which affects up to 25% of children with unrepaired defects [19]. Globally, approximately 80% of the pulmonary hypertension burden occurs in low-resource regions, with CHD representing a major risk factor for pulmonary arterial hypertension, accounting for a substantial proportion of cases in sub-Saharan Africa due to delayed presentation and limited treatment availability [20,21]. Restricted access to diagnostic imaging, cardiac catheterization, and targeted therapies often delays intervention until pulmonary vascular disease has become irreversible, contributing to poorer surgical outcomes and reduced survival following repair [22,23].

Addressing these disparities requires coordinated global health strategies focused on early detection, equitable access to care, and capacity building. Initiatives such as prenatal screening programs, national CHD registries, mobile diagnostic platforms, and telehealth services offer promising opportunities to improve early identification and continuity of care. However, sustainable progress will depend on long-term investment in local infrastructure, workforce training, and collaboration among governments, non-governmental organizations, and academic institutions [24-26].

Advances in diagnostics and surveillance

Recent advances in imaging and biomarker science have improved our ability to detect and monitor pulmonary complications in CHD. High-resolution CT can visualize airway compression, bronchiectasis, and parenchymal abnormalities. Cardiac MRI offers a detailed assessment of ventricular function, vascular anatomy, and flow dynamics. Still widely used, transthoracic echocardiography provides a rapid, noninvasive assessment of both pulmonary pressures and ventricular function, though it has limitations [9].

Biomarkers such as brain natriuretic peptide (BNP, pro-BNP, and NT-pro-BNP) and growth differentiation factor-15 (GDF-15) are increasingly used to identify patients at risk of decompensation. Their utility in longitudinal monitoring and tracking therapeutic responses is under active investigation [1,11]. Nonetheless, significant gaps remain. Longitudinal studies are needed to better define the natural history of CLD in CHD, establish optimal screening intervals, and evaluate emerging therapies in pediatric cohorts.

Toward integrated and preventive care

Managing CLD in CHD requires a multidisciplinary, proactive approach. Early surgical correction of structural defects is essential to preserve lung development. For individuals with established lung disease, personalized therapies targeting airway obstruction, pulmonary vascular disease, or restrictive physiology are crucial.

Preventive strategies must be prioritized:

- Routine pulmonary function testing (age-appropriate).
- Vaccination against respiratory pathogens.
- Nutritional optimization.
- Minimizing exposure to tobacco smoke and environmental pollutants.
- Considering non-invasive ventilatory support in selected cases.

Importantly, psychosocial support should be embedded into care pathways, recognizing the mental health challenges faced by patients and families alike.

Category	Pathophysiology	Clinical Outcomes
Developmental	- Altered fetal pulmonary perfusion during canalicular/saccular stages - Disrupted signaling pathways between heart and lung - Impaired alveolar and vascular maturation	- Pulmonary hypoplasia - Reduced capillary density - Bronchopulmonary dysplasia-like phenotype - Impaired gas exchange and limited reserve
Surgical / Structural	- Non-pulsatile pulmonary blood flow post-Glenn/Fontan - Repeated sternotomies → chest wall deformity, pleural scarring - Phrenic nerve injury → diaphragmatic dysfunction - Airway compression from dilated chambers, grafts, or enlarged pulmonary arteries	- Restrictive lung mechanics - Reduced lung compliance - Ventilation-perfusion mismatch - Airway obstruction or malacia - Decreased exercise capacity
Hemodynamic	- Chronic left-to-right shunts → pulmonary overcirculation - Elevated left atrial/venous pressures - Pulmonary vascular remodeling - Resistance-capacitance (RC) uncoupling	- Pulmonary arterial hypertension - Increased pulmonary vascular resistance - Eisenmenger syndrome - Right ventricular dysfunction and failure
Psychosocial/Functional	- Recurrent hospitalizations and invasive procedures - Chronic hypoxemia and fatigue - Social isolation, missed school, delayed milestones - Anxiety, depression, and body image concerns across development	- Reduced exercise tolerance - Developmental delay - Mood and anxiety disorders (up to 50% prevalence) - Poor quality of life - Family caregiver burden and financial stress

Table 1: *Pulmonary sequelae of congenital heart disease: mechanisms and clinical outcomes.*

Conclusion

Chronic lung disease in congenital heart disease is not merely a comorbidity; it is a core component of the pathophysiological continuum. Addressing it requires shifting from a cardiac-centric to an integrated cardiopulmonary framework. As survival improves, the quality of survival must become the central focus. By embedding pulmonary care across all stages, from fetal diagnosis to adolescence, we can begin to mitigate the long-term burden of CLD and help patients with CHD realize their full potential.

Bibliography

1. Landry LM and CL Jenks. "Pulmonary hypertension associated with congenital heart disease: A clinical primer". *Congenital Heart Disease* 20.3 (2025): 325-339.

2. Ruchonnet-Metrailler I., *et al*. "Pulmonary hypoplasia associated with congenital heart diseases: a fetal study". *PLoS One* 9.4 (2014): e93557.

3. Li DB., *et al*. "Congenital heart disease-associated pulmonary dysplasia and its underlying mechanisms". *American Journal of Physiology-Lung Cellular and Molecular Physiology* 324.2 (2023): L89-L101.

4. Su Z., *et al*. "Improving long-term care and outcomes of congenital heart disease: fulfilling the promise of a healthy life". *Lancet Child and Adolescent Health* 7.7 (2023): 502-518.

5. Abassi H., *et al*. "Impaired pulmonary function and its association with clinical outcomes, exercise capacity and quality of life in children with congenital heart disease". *International Journal of Cardiology* 285 (2019): 86-92.

6. Wacker J., *et al*. "Pulmonary vascular disease as a complication of pediatric congenital heart diseases". *Translational Pediatrics* 12.5 (2023): 1041-1052.

7. Muller J., *et al*. "Number of thoracotomies predicts impairment in lung function and exercise capacity in patients with congenital heart disease". *Journal of Cardiology* 71.1 (2018): 88-92.

8. Georgiev S., *et al*. "Phrenic nerve injury after paediatric heart surgery: is aggressive plication of the diaphragm beneficial?" *European Journal of Cardio-Thoracic Surgery* 44.5 (2013): 808-812.

9. Guerin S., *et al*. "Respiratory morbidity in children with congenital heart disease". *Archives of Pediatrics* 28.7 (2021): 525-529.

10. Jone PN., *et al*. "Pulmonary hypertension in congenital heart disease: A scientific statement from the American Heart Association". *Circulation: Heart Failure* 16.7 (2023): e00080.

11. Sullivan RT., *et al*. "Recent advances in pediatric pulmonary hypertension: Implications for diagnosis and treatment". *Clinical Therapeutics* 45.9 (2023): 901-912.

12. Muneuchi J., *et al*. "Comprehensive assessments of pulmonary circulation in children with pulmonary hypertension associated with congenital heart disease". *Frontiers in Pediatrics* 10 (2022): 1011631.

13. Ladak LA., *et al*. "Health-related quality of life in congenital heart disease surgery in children and young adults: a systematic review and meta-analysis". *Archives of Disease in Childhood* 104.4 (2019): 340-347.

14. Kovacs AH., *et al*. "Psychological outcomes and interventions for individuals with congenital heart disease: a scientific statement from the American heart association". *Circulation: Cardiovascular Quality and Outcomes* 15.8 (2022): e000110.

15. Latal B., *et al*. "Psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease: a systematic review". *BMC Pediatrics* 9 (2009): 6.

16. Marino BS., *et al*. "The impact of neurodevelopmental and psychosocial outcomes on health-related quality of life in survivors of congenital heart disease". *Journal of Pediatrics* 174 (2016): 11-22.e2.

17. Brosig CL., *et al*. "Psychosocial outcomes for preschool children and families after surgery for complex congenital heart disease". *Pediatric Cardiology* 28.4 (2007): 255-262.

18. Grunwald O., *et al*. "Psychiatric and psychological implications of congenital heart disease". *Journal of Clinical Medicine* 14.9 (2025): 3004.

19. Zuhlke L., *et al*. "Congenital heart disease in low- and lower-middle-income countries: Current status and new opportunities". *Current Cardiology Reports* 21.12 (2019): 163.

20. Hasan B., *et al*. "Challenges and special aspects of pulmonary hypertension in middle- to low-income regions: JACC state-of-the-art review". *Journal of the American College of Cardiology* 75.19 (2020): 2463-2477.

21. Hasan BS., *et al*. "Recommendations for developing effective and safe paediatric and congenital heart disease services in low-income and middle-income countries: a public health framework". *BMJ Global Health* 8.5 (2023): e012049.

22. Lai WW., *et al*. "Pediatric and congenital cardiac services: an innovative and empowering approach to global training and equitable care". *Journal of the American Heart Association* 14.8 (2025): e040003.

23. Mocumbi AO., *et al*. "Challenges on the management of congenital heart disease in developing countries". *International Journal of Cardiology* 148.3 (2011): 285-288.

24. Rakha S. "Initiating a fetal cardiac program from scratch in low- and middle-income countries: structure, challenges, and hopes for solutions". *Pediatric Cardiology* 46.2 (2025): 257-266.

25. Zimmerman M and C Sable. "Congenital heart disease in low-and-middle-income countries: Focus on sub-Saharan Africa". *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* 184.1 (2020): 36-46.

26. Ou Y., *et al*. "Prenatal detection of congenital heart diseases using echocardiography: 12-year results of an improving program with 9782 cases". *Frontiers in Public Health* 10 (2022): 886262.

Volume 15 Issue 3 March 2026