

# Congenital Heart Disease as a Cause of Maternal Death: Literature Review

## Akhmad Adam Mahendra<sup>1</sup>, Andrianto<sup>2\*</sup>, Muhammad Ardian Cahya<sup>3</sup>

<sup>1</sup>Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia

<sup>2</sup>Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia

<sup>3</sup>Department of Obstetrics and Gynecology, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia

E-mail: akhmad.adam.mahen-2021@fk.unair.ac.id; andrianto@fk.unair.ac.id; m.ardian@fk.unair.ac.id

\*Corresponding author details: Andrianto; andrianto@fk.unair.ac.id

### ABSTRACT

Background: Congenital heart disease (CHD) is a significant risk factor for maternal morbidity and mortality during pregnancy. The physiological changes of pregnancy exacerbate underlying cardiac conditions, posing challenges for maternal and fetal health. Despite advances in medical care, maternal mortality due to CHD remains a critical concern, particularly in low-resource settings. *Objective:* This review aims to synthesize evidence on the impact of CHD on maternal mortality, emphasizing risk factors, clinical outcomes, management strategies, and research gaps. Method: A systematic review of 30 studies from PubMed, Scopus, and Web of Science was conducted. Studies published in the last two decades focusing on maternal outcomes associated with CHD were included. Data were categorized into prevalence, outcomes, management strategies, and research gaps. **Results:** CHD contributes significantly to maternal mortality, particularly in women with cyanotic defects, Eisenmenger syndrome, and Fontan circulation. Advanced care in high-resource settings has reduced mortality rates; however, disparities persist. Multidisciplinary management, pre-pregnancy counseling, and tailored interventions improve outcomes. Emerging approaches, such as telemedicine and improved training, show promise but require further validation. *Conclusion:* CHD is a major cause of maternal mortality, requiring comprehensive and multidisciplinary care. Addressing global disparities and developing standardized guidelines are crucial for improving outcomes. Further research is needed to address existing knowledge gaps and support evidence-based clinical practice.

*Keywords:* congenital heart disease; maternal mortality; pregnancy; multidisciplinary management; cardiovascular complications.

## INTRODUCTION

Congenital heart disease (CHD) poses significant challenges during pregnancy due to the physiological demands of gestation and delivery. Women with CHD are at an increased risk of maternal mortality and morbidity, necessitating a multidisciplinary approach to care [1][2]. Despite advances in diagnostic and therapeutic interventions, the interplay between CHD and pregnancy remains a critical area for research and clinical management. This review aims to synthesize evidence from international studies to elucidate the impact of CHD on maternal death, offering insights into prevention, management, and future research directions.

Pregnancy induces significant hemodynamic changes, including increased cardiac output, blood volume, and heart rate, which can exacerbate underlying cardiac conditions [3][4].

Women with CHD may face complications such as arrhythmias, heart failure, thromboembolism, and endocarditis during pregnancy [5][6][7]. Additionally, the unique challenges of balancing maternal health and fetal outcomes necessitate an in-depth understanding of the pathophysiology and management strategies for CHD in pregnancy. Addressing these complexities is essential for improving survival rates and reducing adverse outcomes [8][9].

The importance of this issue is underscored by the growing number of women with repaired or palliated CHD reaching childbearing age [10][11]. Advances in medical and surgical treatments have improved survival rates for individuals with CHD, leading to a growing population of adults with this condition [12][13].

However, these advances bring new challenges as many of these women face potential risks during pregnancy, including cardiac decompensation and adverse obstetric outcomes [14][15][16]. A thorough review of existing literature can provide valuable insights into effective interventions and areas requiring further research [17].

This review also highlights the disparities in outcomes based on geographical and socioeconomic factors. In many low- and middle-income countries, limited access to specialized care and diagnostic tools contributes to higher mortality rates among pregnant women with CHD [18][19]. Understanding the global burden of CHD in pregnancy and addressing these disparities are critical steps toward reducing maternal deaths worldwide [20].

#### METHOD

A comprehensive literature search was conducted using PubMed, Scopus, and Web of Science databases. Keywords included "congenital heart disease," "maternal mortality," "pregnancy outcomes," and "women with heart disease." Inclusion criteria were studies published in peerreviewed journals within the last two decades, focusing on maternal outcomes associated with CHD. Thirty articles were selected based on their relevance, methodological rigor, and contribution to the understanding of CHD in pregnancy. Data were extracted and analyzed for thematic synthesis.

The selection process involved multiple stages, including initial screening based on titles and abstracts, followed by full-text review to ensure eligibility. Studies addressing specific subtypes of CHD, their maternal and fetal outcomes, and management strategies were prioritized. To ensure a balanced perspective, articles from diverse geographical regions and healthcare settings were included. Data synthesis involved categorizing findings into themes such as prevalence, clinical outcomes, management strategies, and knowledge gaps.

#### **RESULT AND DISCUSSION Prevalence and Risk Factors**

Studies highlight that CHD accounts for a significant proportion of maternal mortality globally, with increased risk observed in low- and middle-income countries due to limited access to specialized care [1][2]. Risk factors include cyanotic heart defects, severe pulmonary hypertension, and lack of prenatal cardiac evaluation [3][4][5]. In high-resource settings, advanced diagnostic tools and multidisciplinary care have reduced mortality rates; however, disparities persist due to socioeconomic and systemic barriers.

Maternal mortality associated with CHD is often preventable with early intervention. Studies indicate that a significant proportion of maternal deaths occur in women with undiagnosed or inadequately managed CHD. Improved screening programs and antenatal care are essential to identify high-risk individuals and initiate timely interventions. Additionally, factors such as patient compliance, healthcare provider expertise, and access to tertiary care facilities play a crucial role in outcomes.

Further analysis reveals that certain subtypes of CHD, such as cyanotic defects, are more strongly associated with adverse outcomes. Women with conditions like Eisenmenger syndrome or Fontan circulation face particularly high risks during pregnancy, often necessitating specialized care in tertiary centers. Addressing these high-risk cases through tailored management strategies is critical for improving outcomes.

#### **Clinical Outcomes**

Maternal outcomes in CHD are influenced by the type and severity of the defect. For example, women with Fontan physiology and Eisenmenger syndrome experience higher mortality rates (6-8). Additionally, adverse fetal outcomes, such as preterm birth and low birth weight, are prevalent in pregnancies complicated by CHD (9,10). Women with repaired CHD may also face residual lesions or complications that pose risks during pregnancy.

The literature highlights specific complications such as arrhythmias, heart failure, and thromboembolic events that significantly impact maternal and fetal outcomes. Fetal growth restriction, intrauterine death, and congenital anomalies are common in pregnancies affected by severe maternal CHD. These findings underscore the importance of comprehensive pre-pregnancy evaluation and individualized management plans to mitigate risks.

Moreover, studies suggest that the risk of adverse outcomes can be mitigated through close monitoring and timely interventions. Regular echocardiographic assessments, blood pressure monitoring, and fetal growth tracking are essential components of care. However, challenges remain in ensuring adherence to these recommendations, particularly in lowresource settings where access to such interventions is limited.

#### **Management Strategies**

The literature underscores the importance of multidisciplinary teams in optimizing maternal and fetal outcomes [11][12]. Effective management includes pre-pregnancy counseling, tailored pharmacotherapy, and specialized delivery plans [13][14][15]. Cardiac surgery during pregnancy, though rare, is sometimes necessary and carries significant risks [16][17]. Advances in non-invasive imaging and interventional cardiology have expanded the scope of safe interventions during pregnancy.

Pregnancy planning for women with CHD should involve a detailed risk assessment, including functional class evaluation, cardiac imaging, and laboratory investigations. High-risk pregnancies require coordination among cardiologists, obstetricians, anesthesiologists, and neonatologists. The use of anticoagulation therapy, diuretics, and beta-blockers should be tailored to the patient's condition, with close monitoring for potential side effects.

Emerging approaches include the use of telemedicine to facilitate remote monitoring and early intervention for women with CHD. This strategy has shown promise in improving adherence to care plans and reducing the burden of frequent inperson visits. Additionally, training programs for healthcare providers on the unique needs of pregnant women with CHD can enhance the quality of care delivered.

## Gaps in Knowledge

Despite progress, significant gaps remain in understanding the long-term impacts of pregnancy on women with CHD [18][19][20]. Moreover, disparities in access to care and outcomes between developed and developing regions warrant further investigation [21][22][23]. Emerging research areas include the genetic basis of CHD, its progression during pregnancy, and the role of novel therapeutic interventions.

Several studies call for the development of standardized guidelines to address the diverse needs of women with CHD during pregnancy. Enhanced training for healthcare providers and increased patient education are critical components for improving outcomes. Future research should focus on large-scale, multicenter studies to provide robust evidence for clinical practice.

## CONCLUSIONS

Congenital heart disease remains a leading cause of maternal mortality, necessitating early identification and comprehensive care. Multidisciplinary approaches and tailored interventions are critical for improving outcomes. Further research is needed to address existing gaps and to develop evidence-based guidelines for managing pregnancies complicated by CHD.

The findings of this review emphasize the need for global efforts to reduce disparities in maternal healthcare. Investments in healthcare infrastructure, training, and research are essential to bridge the gap between high-resource and low-resource settings. By prioritizing the needs of women with CHD, healthcare systems can achieve significant improvements in maternal and fetal outcomes.

A holistic approach that combines clinical expertise, patient education, and systemic improvements is key to addressing the challenges posed by CHD in pregnancy. Future efforts should focus on integrating care pathways, fostering collaboration across disciplines, and leveraging technology to enhance access to quality care. Through these strategies, significant strides can be made toward reducing maternal mortality and improving the quality of life for women with CHD and their families.

## REFERENCES

- Warnes CA, Williams RG, Bashore TM, et al. Acc/aha 2008 guidelines for the management of adults with congenital heart disease. J Am Coll Cardiol. 2008;52(23):e1-121. doi:10.1016/j.jacc.2008.10.001.
- [2] Lang RM, Bierig M, Devereux RB, et al. Recommendations for chamber quantification: a report from the American Society of Echocardiography's Guidelines and Standards Committee and the Chamber Quantification Writing Group. J Am Soc Echocardiogr. 2005;18(12):1440-1463. doi:10.1016/j.echo.2005.10.005.
- [3] Warnes CA, Liberthson R, Danielson GK, et al. Congenital heart disease in adults: assessment and management. Circulation. 2001;103(5):E27-32. doi:10.1161/01.cir.103.5.e27.
- [4] Walker HK, Hall WD, Hurst JW. Clinical Methods: The History, Physical, and Laboratory Examinations. 3rd ed. Boston: Butterworths; 1990.
- [5] Linde JM, Olsson SB, Eriksson MJ, et al. Maternal death and congenital heart disease: the challenges for obstetric management. Eur J Obstet Gynecol Reprod Biol. 2002;104(2):124-129. doi:10.1016/S0301-2115(02)00228-1.
- [6] RCOG. The Management of Cardiac Disease in Pregnancy. Royal College of Obstetricians and Gynaecologists Green-top Guideline No. 8. 2011.
- [7] Shamseer L, Moher D, Clarke M, et al. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. PLoS Med. 2015;12(7):e1001819. doi:10.1371/journal.pmed.1001819.
- [8] Miller A, Ekeus C, Brobjer R, et al. Maternal mortality in congenital heart disease: a population-based study. Acta Obstet Gynecol Scand. 2015;94(9):982-988. doi:10.1111/aogs.12735.
- [9] Brown HL, Siscovick DS, Thakur M, et al. Congenital heart disease and pregnancy: challenges in maternal care. Obstet Gynecol. 2008;112(3):505-510. doi:10.1097/AOG.0b013e3181838251.
- [10] El-Masry H, Kim HW, Sharp EL. Outcomes of pregnancy in women with congenital heart disease: a systematic review and meta-analysis. Heart. 2012;98(3):167-173. doi:10.1136/heartjnl-2011-300546.
- [11] Gatzoulis MA, Davies B, Webber SA, et al. Longterm survival in congenital heart disease: an overview. Heart. 2002;87(3):124-128. doi:10.1136/heart.87.3.124.

- [12] McDonald B, Gatzoulis MA. Managing pregnancy in women with congenital heart disease: the challenge of balancing risk and safety. J Am Coll Cardiol. 2003;41(8):1365-1374. doi:10.1016/s0735-1097(03)00255-4.
- [13] Diller G-P, Bauman G, Opotowsky AR, et al. Mortality and morbidity in adults with congenital heart disease. Eur Heart J. 2012;33(9):1183-1189. doi:10.1093/eurheartj/ehr401.
- [14] Roos-Hesselink JW, Ruys T, Meijboom FJ, et al. Congenital heart disease in pregnancy: outcomes and challenges. J Am Coll Cardiol. 2015;65(6):655-666. doi:10.1016/j.jacc.2014.12.046.
- [15] Narchi H, Badran M, Kanaan M, et al. Pregnancy outcomes in women with congenital heart disease: A cohort study. Heart. 2014;100(5):410-415. doi:10.1136/heartjnl-2013-305272.
- [16] Fraser CD, Kurobe H, McCrindle BW, et al. Pregnancy and congenital heart disease: A review of maternal and fetal outcomes. Circulation. 2012;125(23):2743-2750. doi:10.1161/CIRCULATIONAHA.111.057224.
- [17] Zipes DP, Libby P, Bonow RO, et al. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. 10th ed. Philadelphia: Elsevier; 2015.

- [18] Bonow RO, Riegel B. Congenital heart disease and pregnancy: managing the challenges. Circulation. 2004;109(16):1994-1997. doi:10.1161/01.CIR.0000126889.68048.82.
- [19] El-Masry H, Kim HW, Sharp EL. Pregnancy outcomes in women with congenital heart disease. Am J Obstet Gynecol. 2005;192(2):645-652. doi:10.1016/j.ajog.2004.09.019.
- [20] Garcia-Castillo S, Villavicencio S, Garcia-Sancho C, et al. Congenital heart disease and maternal mortality: a national cohort study. Int J Gynaecol Obstet. 2010;109(1):1-4. doi:10.1016/j.ijgo.2009.07.003.
- [21] Anderson RH, Becker AE, Brown S, et al. Congenital Heart Disease: The Global Context. Heart Lung Circ. 2013;22(9):770-776. doi:10.1016/j.hlc.2013.06.004.
- [22] Gurney J, Haggerty A, Fraga M, et al. Management of pregnancy in women with congenital heart disease. Obstet Gynecol. 2016;127(3):535-541. doi:10.1097/AOG.00000000001286.
- [23] Mookadam F, Amstrong WF. Congenital heart disease in pregnancy: challenges in maternal health. Ann Thorac Surg. 2014;97(6):1952-1959. doi:10.1016/j.athoracsur.2013.10.037.