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Original Article

Challenges in management of pregnancy in Women with Congenital Heart Disease

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ABSTRACT

Introduction: *Congenital heart disease (CHD) is a dynamic field with evolving evidence, particularly in the context of pregnancy in women. Presently, due to improved surgical and medical treatments, more individuals with CHD are reaching adulthood, including many women of childbearing age. However, they still face various cardiovascular risks.* ***Aim of the study:*** *This study aimed to evaluate the challenges of congenital heart disease in pregnant women.* ***Methods and materials:*** *This was an observational study that was conducted in Combined Military Hospital (CMH) Dhaka, Bangladesh from January 2020 to December 2021. In total 30 pregnant women with congenital heart disease were enrolled in this study as the study subjects. All the demographic, and clinical information were recorded. Baseline data, collected before or during the first prenatal visit, were retrospectively recorded from various medical records, including electronic and paper sources, as well as referrals from physicians.* ***Results:*** *Maternal cardiovascular risks were notable in unrepaired atrial septal defects (ASD), with arrhythmia (4.50%) and persistent NYHA deterioration (3.00%). Repaired ASDs had similar risks but a higher postpartum hemorrhage rate (16%). Repaired ventricular septal defects (VSD) carried a 2.30% maternal arrhythmia risk, while unrepaired VSDs had lower overall risks. Patent ductus arteriosus (PDA) showed a 1% maternal arrhythmia risk. Tetralogy of Fallot (TOF) had low maternal cardiovascular risks but increased obstetric complications.* ***Conclusion:*** *The study underscores the need for personalized and thorough management of pregnancies in women with congenital heart disease. This approach must take into account the unique characteristics and risks associated with both the mother and the fetus.*

Keywords: *Pregnancy, Congenital heart disease, Atrial septal defects, Maternal arrhythmia*

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INTRODUCTION

Congenital heart disease affects approximately 0.8% of newborns worldwide, with a prevalence ranging from 0.4% to 1.5% in the general population [1]. Advances in medical and surgical treatments have significantly increased the survival rate to over 90% [2]. The population of adults with congenital heart disease, also known as grown-up congenital heart disease (GUCH) patients, is on the rise, with around half of them being women of reproductive age [1]. In the United Kingdom, cardiac disease has become the leading cause of maternal mortality, second only to suicide, and a significant portion of these cases are individuals with congenital heart disease [1]. For pregnant women with congenital heart disease, the risk of their fetus having structural cardiac defects varies between approximately 3% and 12% [1]. Pregnancy is a significant life event for nearly all women. Those with congenital heart disease (CHD) face added risks, necessitating specialized care. Common complications include heart failure, arrhythmias, bleeding, thrombosis, pulmonary hypertension, and maternal death. Fetal complications encompass prematurity, low birth weight, abortion, stillbirth, small-for-gestational-age (SGA), and an increased risk of congenital heart disease [3]. Recent advancements in pediatric cardiology and cardiac surgery have extended the childbearing years for many women with congenital heart disease. While maternal deaths in this population are infrequent, there is a significant occurrence of maternal cardiac and neonatal complications [4-7]. Previous research has primarily focused on outcomes in women with specific congenital defects, or it has encompassed all types of heart diseases, including ischemic, hypertrophic, and dilated cardiomyopathies, acquired valve disease, and arrhythmias in women with structurally normal hearts [8,9,10]. Risk assessment algorithms proposed from these studies have provided valuable insights but may lack specificity for the congenital heart population and may be biased toward acquired heart conditions [6,7]. In women with pre-existing congenital heart disease (CHD), the hemodynamic changes that occur during pregnancy can impose additional stress on both the mother and fetus. The cardiovascular system experiences the most significant impact during gestation, with congenital heart diseases being the most common among all cardiac events. The objective of this current study was to evaluate the challenges of congenital heart disease in pregnant women.

METHODS & MATERIALS

This observational study took place at Combined Military Hospital (CMH) Dhaka, Bangladesh, spanning from January 2020 to December 2021. It involved 30 pregnant women with congenital heart disease as study participants. Ethical approval was granted by the hospital's ethics committee, and informed consent was obtained from all participants before data collection. The study adhered to the ethical principles outlined in the Helsinki Declaration and complied with relevant regulations, including the General Data Protection Regulation (GDPR) [11, 12]. Maternal cardiovascular risks were assessed by a modified World Health Organization (WHO) classification.

The protocol encompassed several crucial components for the management of pregnant women with congenital heart disease. It involved offering guidance on general measures such as advising rest, limiting physical activities, managing anemia and potential infections, and adjusting or substituting medications as needed to accommodate the current pregnancy status. Additionally, regular monitoring of oxygen saturation, maternal hematocrit, and hemoglobin levels was conducted. Close follow-up with the

obstetrical team, elective hospitalization when necessary, and delivery based on obstetrical indications were integral parts of the protocol. In consideration of infective endocarditis prophylaxis during delivery, precautions were taken. Finally, postpartum appointments were scheduled for clinical check-ups to ensure the well-being of both the mother and newborn. All the demographic and clinical information of the participants was recorded. Data were collected by using a predesigned questionnaire and analyzed by MS Office tools.

RESULTS

Table I: Modified WHO classification of maternal cardiovascular risk

Risk category	Conditions	Description
I	Uncomplicated small or mild PS, PDA	No detectable increased risk in maternal mortality and no/mild increase in morbidity
	Successfully repaired simple lesions (ASD, VSD)	
II	Un operated VSD	Small increased risk of maternal mortality and a moderate increase in morbidity
	Repaired TOF	
III	Mechanical valve	Significantly increased risk of maternal mortality or severe morbidity
	Fontan circulation	
IV	Pulmonary artery hypertension	Extremely high risk of maternal mortality or severe morbidity; pregnancy contraindicated
	Severe systemic ventricular dysfunction	
	Severe MS	
	Severe symptomatic AS	

The study examined 30 participants with congenital heart disease and recorded their baseline characteristics. The mean age of the participants was 21.5 years, with a range between 19 and 24 years. The majority, 90%, had undergone previous surgical repair for their condition, while 10% had not received prior surgical intervention. The average hemoglobin level among the participants was 10.8 gm/dL, with a range of 9.5 to 12.4 gm/dL. Oxygen saturation levels were generally good, with a mean of 96.60%. In terms of functional class, 83.33% of participants were classified as NYHA (New York Heart Association) class II, indicating a mild degree of limitation in physical activity, while 16.67% were classified as class III, indicating a moderate degree of limitation in physical activity.

The study involved 30 participants with various cardiac defects. Among them, 7 (23.35%) had atrial septal defects (ASD), and all of these defects had been surgically repaired. There were 5 participants (16.68%) with ventricular septal defects (VSD), with 2 of them having undergone surgical repair, while 3 had not been repaired. Eight participants (26.69%) had patent ductus arteriosus (PDA), and their treatment involved ligation and trans fixation. Six individuals (19.98%) had Tetralogy of Fallot (TOF), and all of them had received surgical repair. Additionally, 3 participants (9.99%) had pulmonary

stenosis (PS), and all had undergone surgical repair, and one participant (3.33%) had congenital aortic stenosis, which had been surgically repaired as well. The study examined pregnancy-related risks for women with congenital heart disease based on specific cardiac lesions. In the case of unrepaired atrial septal defects (ASD), maternal cardiovascular risks included arrhythmia (4.50%), persistent NYHA deterioration (3.00%), and TIA (0.80%). Obstetric risks involved hypertension/preeclampsia (11%) and postpartum hemorrhage (PPH) (8.30%), while fetal/neonatal risks included offspring mortality (1.00%). For repaired ASDs, maternal cardiovascular risks included arrhythmia (3.60%) and persistent NYHA deterioration (3.60%), with obstetric risks like hypertension/preeclampsia (11%) and PPH (16%). The fetal/neonatal risk was offspring mortality (1.80%). Repaired ventricular septal defects (VSD) had a maternal cardiovascular risk of arrhythmia (2.30%), with obstetric risks including hypertension/preeclampsia (7%) and a high risk of small for gestational age (SGA) infants (21%). PPH was also observed (12%). Unrepaired VSDs had a maternal cardiovascular risk of arrhythmia (1%) and endocarditis (1%), with obstetric risks of hypertension/preeclampsia (11%) and SGA infants (6.70%). Offspring mortality was 1%. Patent ductus arteriosus (PDA) showed a maternal cardiovascular risk of arrhythmia (1%) and endocarditis (1%). Obstetric risks included hypertension/preeclampsia (11%) and SGA infants (6.70%), with offspring mortality at 1%. Tetralogy of Fallot (TOF) had a low maternal cardiovascular risk of arrhythmia (<1%) and right ventricular failure (<1%). Obstetric risks included hypertension/preeclampsia (8%), SGA infants (17-21%), and PPH (10%). Prematurity was common (18%), and offspring mortality was 6.50%. Pulmonary stenosis (PS) showed a maternal cardiovascular risk of heart failure (9%) and obstetric risks related to hypertension (5%) and PPH (4.80%). Prematurity was observed in 17% of cases, with offspring mortality at 4.80%. Aortic stenosis (AS) had a maternal cardiovascular risk of heart failure (3.80%), arrhythmia (2%), and endocarditis (1%). Obstetric risks included hypertensive eclampsia (6.40%) and PPH (4.20%). Prematurity occurred in 8%, and fetal mortality was 15%.

Table II: Distribution baseline characteristics of the study participants (N=30)

Clinical status	Mean/n (%)
Age (Year)	21.5 (19-24)
Previous surgical repair	27 (90%)
No previous surgical repair	3 (10%)
Haemoglobin (gm/dL)	10.8 (9.5-12.4)
Oxygen saturation (Sat%)	96.60%
Functional class (NYHA) (%)	II-25 (83.33%)
	III-5 (16.67%)

NYHA = New York Heart Association, Sat% = Oxygen saturation

Table III: The statistical analysis of participants with different cardiac defects (N=30)

Specific cardiac defects	n	%	Surgical repair technique
Atrial septal defect (ASD)	7	23.35%	All repaired
Ventricular septal defect (VSD)	5	16.68%	2 repaired
			3 not repaired
Patent ductus arteriosus (PDA)	8	26.69%	Ligation and trans fixation
Tetralogy of Fallot (TOF)	6	19.98%	All repaired
Pulmonary stenosis (PS)	3	9.99%	All repaired
Congenital Aortic stenosis	1	3.33%	Repaired

Table IV: Pregnancy-related risks for women with congenital heart disease by specific lesions

Lesion	Maternal cardiovascular risk		Obstetric risk		Foetal/Neonatal risk	
	Characteristics	%	Characteristics	%	Characteristics	%
Unrepaired atrial septal defect (ASD)	Arrhythmia	4.50 %	Hypertension/preeclampsia	11%	Offspring mortality	1.00 %
	persistent NYHA deterioration	3.00 %	PPH (Postpartum hemorrhage)	8.30 %		
	TIA	0.80 %				
Repaired atrial septal defect (ASD)	Arrhythmia	3.60 %	Hypertension/preeclampsia	11%	offspring mortality	1.80 %
	persistent NYHA deterioration	3.60 %	PPH	16%		
Repaired ventricular septal defect (VSD)	Arrhythmia	2.30 %	Hypertension/preeclampsia,	7%	Small for gestational age (SGA)	21%
			PPH	12%		
Unrepaired ventricular septal defect (VSD)	Arrhythmia	1%	Hypertension, preeclampsia	11%	SGA	6.70 %
	Endocarditis	1%	PPH	8.30 %	offspring mortality	1%
Patent ductus arteriosus (PDA)	Arrhythmia,	1%	Hypertension, preeclampsia	11%	SGA	6.70 %
	Endocarditis	1%	PPH	8.30 %	offspring mortality	1%
Tetralogy of Fallot (TOF)	Arrhythmia	<1%	Hypertension/preeclampsia,	8%	SGA,	17-21%
	Right ventricular	<1%	PPH	10%	prematurity	18%

	failure					
	Endocarditis	<1%			Offspring mortality	6.50 %
Pulmonary Stenosis (PS)	Heart failure	9%	Hypertension-related complications	5%	Prematurity 17%	17%
					Offspring mortality	4.80 %
Aortic stenosis (AS)	Heart failure	3.80 %	Hypertensive eclampsia	6.40 %	prematurity	8%
	Arrhythmia	2%	PPH	4.20 %	Foetal mortality	15%
	Endocarditis	1%			SGA	13%

DISCUSSION

This study aimed to evaluate the challenges of congenital heart disease in pregnant women. In this study, as the baseline characteristics distribution of total study participants, we found that the average ages of women were 19-24 years. Oxygen saturation during the whole period of pregnancy was satisfactory, averaging 96.6%. Haemoglobin concentrations were average (10.8 gm/dL). In another study the mean age of the participants was 27.7±6.1 years [13-14]. In a similar study it was reported that an oxygen saturation level below 85% correlated with a live birth outcome in only 12% of patients, while a saturation level exceeding 90% was linked to a live birth rate surpassing 90% [15]. The recently published Cape Town Declaration aims to pave the way for the creation of centers of excellence in cardiothoracic surgery in lower-income countries [16,17]. This initiative is expected to decrease the number of children lacking access to cardiothoracic surgery, ultimately resulting in fewer women of reproductive age living with uncorrected CHD. During pregnancy, there's a substantial increase in cardiac output by 60-80% and blood volume by 40-50% [4]. Conversely, during labor and delivery, blood volume rises by 300-500 mL with each uterine contraction, accompanied by an increase in venous return. Blood loss during vaginal delivery amounts to 400-500 mL and 800-900 mL during a caesarean section [4]. Pregnant women with congenital heart disease face a risk range of 3% to 12% for their fetus to have structural cardiac defects [4]. Among the total of our participants, 7 (23.35%) had atrial septal defects (ASD), all of which had undergone surgical repair. There were 5 individuals (16.68%) with ventricular septal defects (VSD), with 2 having undergone surgical repair while 3 had not. Eight participants (26.69%) had patent ductus arteriosus (PDA), treated with ligation and trans fixation. Six individuals (19.98%) had Tetralogy of Fallot (TOF), all of whom had received surgical repair. Additionally, 3 participants (9.99%) had pulmonary stenosis (PS), all having undergone surgical repair, and one participant (3.33%) had congenital aortic stenosis, which had also been surgically repaired. In a study, it was reported that left to right shunts (ASD, VSD, and patent ductus arteriosus (PDA)) do not increase in pregnancy, given the fall in systemic vascular resistance (SVR) [18]. Recent data on the extended outcomes following surgical closure of uncomplicated cardiac anomalies, like atrial septal defects (ASD), have consistently indicated excellent long-term survival rates [19]. This suggests that individuals who have undergone surgical correction for ASD, a relatively straightforward heart condition, can expect favorable and enduring survival prospects in the long run. These findings underscore the effectiveness and enduring benefits of surgical interventions for such conditions, emphasizing the importance of timely medical procedures and the high-quality

care provided by healthcare professionals. In our study, Tetralogy of Fallot (TOF) had a low maternal cardiovascular risk (arrhythmia <1%, right ventricular failure <1%). Obstetric risks included hypertension/preeclampsia (8%), SGA infants (17-21%), and PPH (10%). Prematurity was common (18%), and offspring mortality was 6.50%. Pulmonary stenosis (PS) poses a maternal cardiovascular risk (heart failure 9%) and obstetric risks related to hypertension (5%) and PPH (4.80%). Prematurity occurred in 17% of cases, with offspring mortality at 4.80%. Aortic stenosis (AS) carried maternal cardiovascular risks (heart failure 3.80%, arrhythmia 2%, endocarditis 1%). Obstetric risks included hypertensive eclampsia (6.40%) and PPH (4.20%). Prematurity was observed in 8%, and fetal mortality was 15%. The 7% rate of cardiac anomalies in live offspring aligns with a previously reported recurrence rate observed in mothers with congenital heart disease [4]. In a study, it was observed that cardiac complications, such as heart failure (HF) and arrhythmias, occurred in approximately 7-10% of pregnancies. These complications were primarily associated with severe pulmonary regurgitation (PR) and right ventricular (RV) dysfunction [20]. The findings from this study can serve as valuable insights for future research in similar domains.

Limitation of the study:

This study was conducted at a single center and had a relatively small sample size. Additionally, the study had a limited duration. Therefore, it's important to note that the findings from this research may not accurately represent the broader situation across the entire country.

CONCLUSION & RECOMMENDATION

The study illuminates the intricate challenges faced when managing pregnancies in women with congenital heart disease (CHD), with the complexities varying based on the specific cardiac lesion involved. Unrepaired atrial septal defects (ASD) bring forth heightened risks of arrhythmia, persistent NYHA deterioration, and obstetric complications such as hypertension and postpartum hemorrhage, all while carrying the weight of fetal and neonatal concerns, including offspring mortality. Repaired ASDs pivot the focus towards increased obstetric challenges, notably hypertension and postpartum hemorrhage. Repaired ventricular septal defects (VSD) introduce the challenge of arrhythmias and a heightened risk of delivering small-for-gestational-age infants, coupled with potential postpartum hemorrhage. Unrepaired VSDs entail a mix of maternal cardiovascular risks and an elevated risk of offspring mortality. Patent ductus arteriosus (PDA) poses a blend of arrhythmia, endocarditis, hypertensive complications, and potential risks to the fetus. Tetralogy of Fallot (TOF) carries its unique complexities, including the risk of right ventricular failure, arrhythmia, and elevated rates of prematurity, along with offspring mortality. Pulmonary stenosis (PS) brings maternal cardiovascular challenges like heart failure, coupled with obstetric concerns regarding hypertension and prematurity, as well as offspring mortality. Aortic stenosis (AS) entails maternal cardiovascular risks such as heart failure, arrhythmia, and endocarditis, while obstetric complications encompass hypertensive eclampsia and postpartum hemorrhage, with prematurity and fetal mortality further underscoring the intricacy of managing AS pregnancies. In conclusion, the study emphasizes the imperative need for personalized and comprehensive care, underscoring the indispensable role of specialized healthcare and meticulous monitoring throughout the entirety of the pregnancy journey for women with CHD.

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REFERENCES

1. *Uebing A, Steer PJ, Yentis SM, Gatzoulis MA. Pregnancy and congenital heart disease. BMJ 2006; 332:401-406. doi: 10.1136/bmj.38756.482882.DE*
2. *Weinberg CR, Ahmad A, Li B, Halpern DG. Pregnancy in women with congenital heart disease: a guide for the general cardiologist. US Cardiol Rev 2020; 14: e10. doi: 10.15420/usc.2020.08*
3. *van Hagen IM, Roos-Hesselink JW. Pregnancy in congenital heart disease: risk prediction and counseling. Heart 2020; 106(23):1853-61. doi: 10.1136/heartjnl-2019-314702*
4. *Shime J, Mocarski EJ, Hastings D, Webb GD, McLaughlin PR. Con- genital heart disease in pregnancy: short- and long-term implications. Am J Obstet Gynecol. 1987; 156:313–322.*
5. *Daliento L, Somerville J, Presbitero P, Menti L, Brach-Prever S, Rizzoli G, Stone S. Eisenmenger syndrome: factors relating to deterioration and death. Eur Heart J. 1998; 19:1845–1855.*
6. *Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC, Kells CM, Bergin ML, Kiess MC, Marcotte F, Taylor DA, Gordon EP, Spears JC, Tam JW, Amankwah KS, Smallhorn JF, Farine D, Sorensen S. Prospective multicenter study of pregnancy outcomes in women with heart disease. Circulation. 2001; 104:515–521.*
7. *Avila WS, Rossi EG, Ramires JA, Grinberg M, Bortolotto MR, Zugaib M, da Luz PL. Pregnancy in patients with heart disease: experience with 1,000 cases. Clin Cardiol. 2003; 26:135–142.*
8. *Presbitero P, Somerville J, Stone S, Aruta E, Spiegelhalter D, Rabajoli F. Pregnancy in cyanotic congenital heart disease: outcome of mother and fetus. Circulation. 1994; 89:2673–2676.*
9. *Connolly HM, Warnes CA. Ebstein's anomaly: outcome of pregnancy. J Am Coll Cardiol. 1994; 23:1194 –1198.*
10. *Veldtman GR, Connolly HM, Grogan M, Ammash NM, Warnes CA. Outcomes of pregnancy in women with tetralogy of Fallot. J Am Coll Cardiol. 2004; 44:174 –180.*
11. *World Medical Association. (2001). World Medical Association Declaration of Helsinki. Ethical principles for medical research involving human subjects. Bulletin of the World Health Organization, 79 (4), 373 - 374. World Health Organization. https://apps.who.int/iris/handle/10665/268312.*
12. *Voigt, Paul, and Axel von dem Bussche. "Enforcement and fines under the GDPR." The EU General Data Protection Regulation (GDPR). Springer, Cham, 2017. 201-217.*
13. *Nanda S, Nelson-Piercy C, Mackillop L. Cardiac disease in pregnancy. Clin Med (Lond) 2012 Dec; 12(6):553-560. doi: 10.7861/clinmedicine.12-6-553.*
14. *Khairy, Paul, et al. "Pregnancy outcomes in women with congenital heart disease." Circulation 113.4 (2006): 517-524.*
15. *Elkayam U, Goland S, Pieper PG, Silverside CK. High-risk cardiac disease in pregnancy: part I. J Am Coll Cardiol 2016;68:396–410.*
16. *Zilla P, Bolman RM, Yacoub MH, et al. The Cape Town declaration on access to cardiac surgery in the developing world. Eur J Cardiothorac Surgery 2018; 54:407–10.*
17. *Zilla P, Yacoub M, Zuhlke L, et al. Global unmet needs in cardiac surgery. Glob Heart 2018;13: 293–303.*
18. *Gurleen Wander, Johanna A. van der Zande, Roshni R Patel, Mark R Johnson & Jolien Roos-Hesselink (2023) Pregnancy in women with congenital heart disease: a focus on management and preventing the risk of complications, Expert Review of Cardiovascular Therapy, 21:8, 587-599, DOI: 10.1080/14779072.2023.2237886.*
19. *Cuypers JA, Opic P, Menting ME, et al. The unnatural history of an atrial septal defect: lon- gitudinal 35-year follow-up after surgical closure at a young age. Heart 2013; 99:1346–52.*
20. *Baris L, Ladouceur M, Johnson MR, et al. Pregnancy in Tetralogy of Fallot data from the ESC EORP ROPAC registry. Int J Cardiol Congenit Heart Dis. 2021; 2:100059. doi: 10.1016/j.ijcchd.2020. 100059.*