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AGAINST ALL ODDS: REPEATED SUCCESSFUL PREGNANCIES IN TWO WOMEN WITH UNREPAIRED SINGLE-VENTRICLE PHYSIOLOGY

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ABSTRACT

Introduction: Pregnancy in women with unrepaired single-ventricle physiology is rare and associated with substantial maternal and foetal risk. Evidence regarding pregnancy outcomes in this population remains limited.

Case description: We report three successful pregnancies in two women with unrepaired double-inlet left ventricle, transposition of the great arteries and valvular/sub-valvular pulmonary stenosis. Both women were classified as modified World Health Organization class IV and received multidisciplinary care throughout pregnancy. Despite complex congenital heart disease, systemic ventricular function and oxygen saturation remained stable. All three pregnancies resulted in live births. However, foetal growth restriction, oligohydramnios, placental insufficiency, prematurity and prolonged neonatal hospitalisation occurred in all pregnancies. One patient successfully completed two consecutive pregnancies without maternal cardiovascular deterioration.

Discussion: Pulmonary outflow obstruction, relatively preserved oxygen saturation and stable systemic ventricular function may have contributed to maternal haemodynamic stability. However, substantial foetal and neonatal morbidity in all three pregnancies underscores that favourable maternal outcomes do not establish pregnancy safety in this population.

Conclusion: Carefully selected women with unrepaired single-ventricle physiology and balanced circulation may tolerate pregnancy better than expected, although foetal complications remain common. Multidisciplinary management in specialised congenital heart disease centres is essential.

KEYWORDS

Single ventricle, double-inlet left ventricle, pulmonary stenosis

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LEARNING POINTS

- Successful pregnancy may be possible in selected women with unrepaired single-ventricle physiology and balanced circulation.
- Favourable maternal outcomes do not preclude foetal complications, including foetal growth restriction, prematurity and prolonged neonatal hospitalisation.
- Multidisciplinary management in specialised congenital heart disease centres is essential.

INTRODUCTION

Congenital heart disease (CHD) affects approximately 1% of live births worldwide, and advances in medical and surgical care have enabled increasing numbers of patients with complex CHD to survive into adulthood and reach reproductive age[1,2]. Single-ventricle physiology represents one of the most complex forms of CHD and encompasses a heterogeneous group of rare malformations, including hypoplastic left heart syndrome, tricuspid atresia and double-inlet ventricle[3].

Pregnancy in women with single-ventricle physiology carries substantial maternal and foetal risk because the physiological increases in blood volume and cardiac output may be poorly tolerated by a solitary systemic ventricle[4]. According to current European Society of Cardiology guidelines, maternal risk depends on functional status, ventricular function, cyanosis, valvular dysfunction, arrhythmias and other complications. Women with severe ventricular dysfunction or New York Heart Association class III–IV symptoms may be classified as modified World Health Organization class IV, in which pregnancy is generally discouraged[4].

Despite these risks, successful pregnancies have been reported in women with unrepaired single-ventricle physiology, although the available evidence remains limited to case reports and small case series[5-7]. We describe three pregnancies resulting in live births in two women with an unrepaired double-inlet left ventricle, transposition of the great arteries and valvular/sub-valvular pulmonary stenosis.

CASE DESCRIPTION

Case 1

A 28-year-old primigravida with unrepaired cyanotic CHD was referred at 30 weeks of gestation. Double-inlet left ventricle (DILV) with transposition of the great arteries (TGA) and valvular/sub-valvular pulmonary stenosis had been diagnosed at 19 years of age. Before pregnancy, her resting oxygen saturation was 88–92%. She was classified as modified World Health Organization risk class IV because of New York Heart Association class III symptoms. Following multidisciplinary counselling regarding the substantial maternal and foetal risks, she elected to continue the pregnancy and was followed by cardiology, maternal-foetal medicine, cardiac surgery and anaesthesia teams.

At presentation she was clinically stable, with an oxygen saturation of 88–92% on room air. Haemoglobin remained

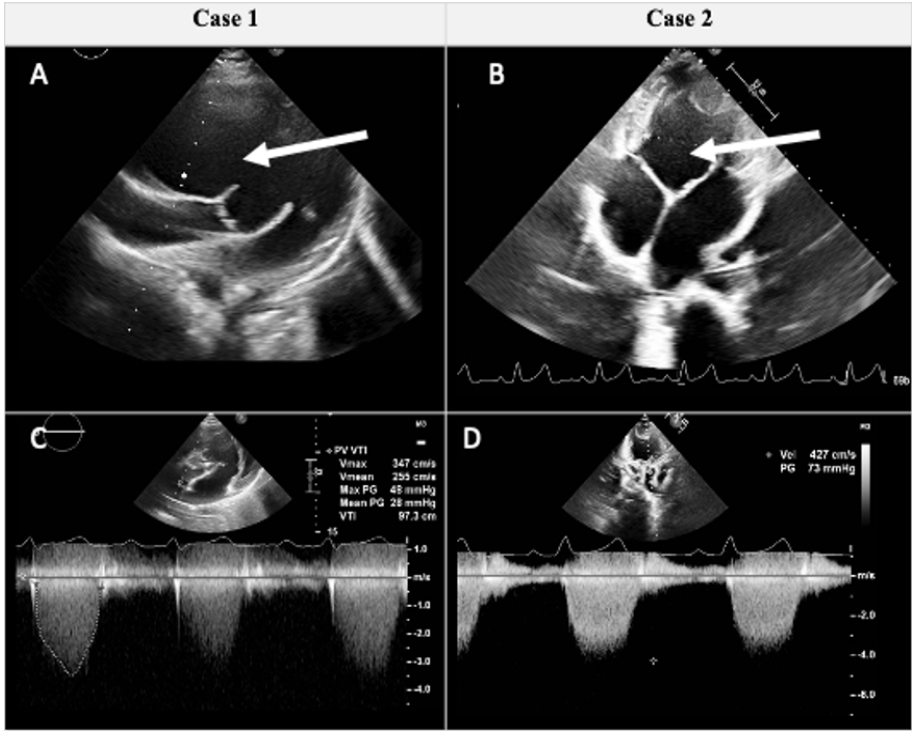


Figure 1. *Echocardiographic findings in two women with unrepaired double-inlet left ventricle, transposition of the great arteries and pulmonary stenosis. A, B) Two-dimensional echocardiographic images demonstrating double-inlet left ventricular morphology. White arrows indicate the dominant morphologic left ventricle; C, D) Continuous-wave Doppler interrogation of the pulmonary outflow tract demonstrating significant valvular/sub-valvular pulmonary stenosis with preserved antegrade pulmonary blood flow. The peak instantaneous gradients were 48 mmHg and 73 mmHg in cases 1 and 2, respectively.*

Variable	Case 1	Case 2
Age	28	27
Gravidity	G1	G1
Cardiac anatomy	DILV with TGA, valvular/sub-valvular pulmonary stenosis	DILV with TGA, valvular/sub-valvular pulmonary stenosis
Oxygen saturation (%)	88–92	88–92
NYHA class	III	III
Haemoglobin (g/dl)	12–14	12.5–12.6
Systemic ejection fraction (%)	40–42	56–58
Systemic ventricular end-diastolic diameter (cm)	8.3	8.1
Peak pulmonary stenosis gradient (mmHg)	48–64	64–75
Mitral regurgitation	Moderate	Mild–moderate
Tricuspid regurgitation	Mild–moderate	Mild
Systemic ventricular tissue Doppler S′ velocity (cm/s)	6.9–9	7.6
dP/dt (mmHg/s)	478–649	909–1,010

Table 1. *Baseline clinical and echocardiographic characteristics of two women with double-inlet left ventricle, transposition of the great arteries and pulmonary stenosis.*

Abbreviations: DILV, double-inlet left ventricle; NYHA, New York Heart Association functional class; TGA, transposition of the great arteries; dP/dt, rate of ventricular pressure rise; Doppler S′, peak systolic tissue Doppler velocity.

between 12 and 14 g/dl throughout pregnancy. Serial echocardiography demonstrated DILV with TGA and moderate valvular/sub-valvular pulmonary stenosis *(Fig. 1)*. Systemic ventricular systolic function remained stable, with an ejection fraction of 40–42% and pulmonary stenosis gradients of 48–64 mmHg. Tissue Doppler S′ velocity ranged from 6.9 to 9 cm/s, and dP/dt ranged from 478 to 649 mmHg/s. Moderate mitral regurgitation and mild-to-moderate tricuspid regurgitation were present without significant deterioration *(Table 1)*.

Foetal anatomical assessment at 16 weeks showed no structural abnormalities. At 28 weeks, foetal ultrasonography demonstrated early foetal growth restriction and oligohydramnios, although uteroplacental and foetal Doppler findings remained normal.

Because of foetal growth restriction and oligohydramnios in the setting of severe maternal cardiac disease, caesarean delivery under regional anaesthesia was performed at 30 weeks and 4 days. A male infant weighing 1,200 g was delivered with Apgar scores of 5 and 6 at 1 and 5 minutes, respectively. The infant required neonatal intensive care for highly preterm birth and respiratory distress syndrome but was discharged without supplemental oxygen after 44 days. The maternal peripartum course was uncomplicated, without heart failure, sustained arrhythmia or other major cardiovascular events.

Case 2

A 27-year-old woman with unrepaired complex CHD was referred to a tertiary centre at 15 weeks of her first pregnancy. DILV with TGA and combined valvular/sub-valvular pulmonary stenosis had been diagnosed at birth. Cardiac catheterisation during childhood confirmed a dominant left-sided single ventricle, a large ventricular septal communication, TGA and moderate-to-severe pulmonary stenosis. Surgical palliation had been recommended on several occasions but was not undertaken.

At presentation she was haemodynamically stable, with a blood pressure of 132/78 mmHg, heart rate of 84 beats/minute and oxygen saturation of 90% on room air. Physical examination demonstrated mild acrocyanosis and a harsh systolic murmur without peripheral oedema. Serial NT-proBNP concentrations ranged from 77.8 to 314.5 pg/ml. Despite the transient increase, she had no clinical evidence of heart failure or need for escalation of cardiac therapy. Following multidisciplinary assessment, she was classified as modified World Health Organization risk class IV. After counselling regarding the substantial maternal and foetal risks and the option of pregnancy termination, she elected to continue the pregnancy.

Throughout gestation she remained clinically stable, with exertional dyspnoea and resting oxygen saturation of 88–92%. Serial echocardiography demonstrated DILV with TGA, moderate-to-severe valvular/sub-valvular pulmonary stenosis, preserved systemic ventricular systolic function with an ejection fraction of 56–58% and moderate atrioventricular valve regurgitation without significant deterioration *(Table 1 and Fig. 1)*.

Progressive foetal growth restriction, oligohydramnios and placental insufficiency developed during the third trimester. Caesarean delivery under regional anaesthesia was performed at 31 weeks and 2 days. A female infant weighing 1,313 g was delivered with Apgar scores of 5 and 6 at 1 and 5 minutes, respectively. The infant required neonatal intensive care for a highly preterm birth, respiratory distress syndrome, sepsis and bronchopulmonary dysplasia but was discharged without supplemental oxygen after 52 days. The maternal

Variable	Case 1 Pregnancy	Case 2 Pregnancy 1	Case 2 Pregnancy 2
Pregnancy risk class (mWHO)	IV	IV	IV
Major pregnancy complications	Foetal growth restriction, oligohydramnios	Foetal growth restriction, oligohydramnios	Foetal growth restriction, chronic placental insufficiency
Gestational age at delivery	30 weeks + 4 days	31 weeks + 2 days	31 weeks
Mode of delivery	Caesarean section	Caesarean section	Repeat caesarean section
Mode of anaesthesia	Regional	Regional	Regional
Neonatal sex	Male	Female	Male
Birth weight (g)	1,200	1,313	1,660
Apgar score	5/6	5/6	6/7
Duration of neonatal hospitalisation (days)	44	52	36
Maternal survival	Yes	Yes	Yes
Neonatal survival	Yes	Yes	Yes

Table 2. *Pregnancy, delivery and neonatal outcomes in three pregnancies from two women with unrepaired single-ventricle physiology. Abbreviations: mWHO, modified World Health Organization – a maternal cardiovascular risk classification.*

peripartum course was uncomplicated.

Two years later, the patient conceived again. She remained clinically stable without overt heart failure, and serial echocardiography showed no significant change in systemic ventricular function, atrioventricular valve regurgitation or pulmonary stenosis *(Table 2 and Fig. 1)*. Foetal growth restriction and placental insufficiency recurred. Repeat caesarean delivery under regional anaesthesia was performed at 31 weeks. A male infant weighing 1,660 g was delivered with Apgar scores of 6 and 7. The infant required prolonged neonatal intensive care for respiratory distress and pneumonia but was discharged after 36 days. Maternal recovery was uncomplicated, and tubal ligation was performed with the patient's consent. Subsequent cardiology assessment demonstrated stable ventricular function without clinical decompensation.

DISCUSSION

We describe three pregnancies in two women with unrepaired single-ventricle physiology, including two consecutive pregnancies in one woman, without major maternal cardiovascular deterioration. Both women had a DILV with TGA and significant valvular/ sub-valvular pulmonary stenosis.

The natural history of an unrepaired single ventricle depends substantially on pulmonary blood flow. Unrestricted pulmonary blood flow may cause pulmonary over-circulation, ventricular volume overload, heart failure and pulmonary vascular disease. Conversely, excessive pulmonary outflow obstruction causes severe cyanosis. Rarely, moderate pulmonary stenosis may provide a relatively balanced circulation by limiting pulmonary over-circulation while maintaining sufficient pulmonary blood flow[2,3,8]. This physiology may have contributed to our patients' survival into adulthood and their haemodynamic stability during pregnancy. However, without contemporary invasive haemodynamic assessment, balanced circulation and the absence of significant pulmonary vascular disease cannot be definitively established and are inferred from anatomical findings, pulmonary stenosis gradients, oxygen saturation and stable ventricular function.

Both women maintained resting oxygen saturation of 88–92%. Previous observations in cyanotic CHD indicate that foetal outcomes deteriorate markedly when maternal oxygen saturation is below 85%[9]. Their relatively preserved oxygen saturation, stable systemic ventricular function and absence of overt heart failure or sustained arrhythmia may have contributed to the favourable maternal outcomes. Nevertheless, these findings should not be interpreted as evidence that pregnancy is safe in women with unrepaired single-ventricle physiology.

In contrast to the maternal courses, foetal and neonatal morbidity was substantial. Foetal growth restriction occurred in all three pregnancies, with oligohydramnios or placental insufficiency leading to highly preterm delivery at approximately 31 weeks. All three infants had low birth weight and required prolonged neonatal intensive care. These outcomes are consistent with previous reports of high rates of foetal growth restriction, prematurity and low birth weight among women with cyanotic CHD and uncorrected single-ventricle physiology[5-7,9].

One woman completed two pregnancies without apparent maternal cardiovascular deterioration. Although repeated pregnancies in women with an unrepaired single ventricle and TGA have previously been reported[10], published experience remains limited to isolated reports and small case series[5-7,10]. Our observations suggest that some women with pulmonary outflow obstruction, acceptable oxygen saturation and stable ventricular function may tolerate pregnancy better than anticipated. However, individual favourable outcomes do not override the very high guideline-based maternal risk or justify recommending

pregnancy in this population[4].

Current guidelines recommend vaginal delivery for most women with cardiovascular disease because it is generally associated with less blood loss and lower thromboembolic and infectious risks than caesarean delivery[4]. In our patients, all three caesarean deliveries were undertaken for foetal growth restriction, oligohydramnios or placental insufficiency rather than maternal cardiovascular deterioration. We used regional anaesthesia with careful attention to preload, systemic vascular resistance, ventricular function and avoidance of increased pulmonary vascular resistance.

CONCLUSION

Favourable maternal outcomes are possible in selected women with unrepaired single-ventricle physiology and relatively balanced pulmonary blood flow. However, foetal growth restriction, highly preterm delivery and prolonged neonatal hospitalisation occurred in all three pregnancies. Pregnancy in this population remains very high risk and requires individualised counselling and multidisciplinary management in a tertiary centre with expertise in adult CHD and maternal-foetal medicine.

REFERENCES

1. Xu J, Li Q, Deng L, Xiong J, Cheng Z, Ye C. Global, regional, and national epidemiology of congenital heart disease in children from 1990 to 2021. Front Cardiovasc Med. 2025;12:1522644. doi: 10.3389/fcvm.2025.1522644.
2. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021;42(6):563-645. doi: 10.1093/eurheartj/ehaa554.
3. Rao PS. Single Ventricle-A Comprehensive Review. Children (Basel). 2021;8(6):441. doi: 10.3390/children8060441.
4. De Backer J, Haugaa KH, Hasselberg NE, de Hosson M, Brida M, Castelletti S, et al. 2025 ESC Guidelines for the management of cardiovascular disease and pregnancy. Eur Heart J. 2025;46(43):4462-4568. doi: 10.1093/eurheartj/ehaf193. Erratum in: Eur Heart J. 2026;47(7):812. doi: 10.1093/eurheartj/ehaf1011.
5. Nilofer AR, Zaki SA. Successful pregnancy in a patient with univentricular heart and pulmonary stenosis. Heart Views. 2012;13(2):71-3. doi: 10.4103/1995-705X.99232.
6. Theodoridis TD, Anagnostou E, Zepiridis L, Dinas K, Bontis J. Successful pregnancy and caesarean section delivery in a patient with single ventricle and transposition of the great arteries. J Obstet Gynaecol. 2005;25(1):69-70. doi: 10.1080/01443610400025911.
7. Keepanasseril A, Raj A, Pillai AA, Baghel J, Plakkal N, Satheesh S. Pregnancy outcome in women with an uncorrected single ventricle: A single-centre experience from South India. Obstet Med. 2020;13(3):137-141. doi: 10.1177/1753495X19845822.
8. Poterucha JT, Anavekar NS, Egbe AC, Julsrud PR, Connolly HM, Ammash NM, et al. Survival and outcomes of patients with unoperated single ventricle. Heart. 2016;102(3):216-22. doi: 10.1136/heartjnl-2015-308440.
9. 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(6):2673-6. doi: 10.1161/01.cir.89.6.2673.
10. Leibbrandt G, Münch U, Gander M. Two successful pregnancies in a patient with single ventricle and transposition of the great arteries. Int J Cardiol. 1982;1(3-4):257-62. doi: 10.1016/0167-5273(82)90087-0.