Anesthetic management for cesarean section in parturient with an uncorrected single ventricle: A case report and literature review

Yu Du, PhD, Yingzi Yang-Liu, MD, Bin Chen, MD, Ji Wang, PhD

Abstract

Rationale: Patients with a single ventricle, who have not undergone surgery, reportedly have a lower survival rate. Furthermore, multiple pregnancies are rare among these females. We reported a case of anesthesia management of cesarean section in an uncorrected single-ventricular multi-pregnancy woman and review the anesthesia management of the published similar cases.

Patient concerns: An uncorrected single ventricular pregnant woman with a cardiac function of New York Heart Association class II, who had experienced one spontaneous abortion and three vaginal deliveries, was scheduled for cesarean section at 37+6 weeks of gestation.

Diagnoses: Echocardiography revealed a complex congenital heart disease in the mother: a single ventricle (the left ventricle is dominant), atroventricular valve ectopic, double-inlet left ventricle, abnormal location of the great arteries, probably pulmonary stenosis, atrial septal defect, and left-to-right shunt. The fetus was in breech presentation with umbilical cord around the neck.

Interventions: Cesarean section was successfully performed under the combined spinal epidural anesthesia with careful monitoring.

Outcomes: Both mother and newborn recovered good and were discharged from the hospital 5 days after surgery without any adverse reactions.

Lessons: Single ventricular pregnant woman with a cardiac function of New York Heart Association class I–II could tolerate pregnancy and delivery well. Both general and regional anesthesia are applicable to cesarean section in these patients. The principle of anesthesia management is to maintain the appropriate balance between systemic vascular resistance and pulmonary vascular resistance, as well as to maintain preload and cardiac output.

Abbreviations: ABG = arterial blood gas analysis, BP = blood pressure, CHD = congenital heart disease, CO = cardiac output, HR = heart rate, PaCO2 = partial pressure of carbon dioxide, PaO2 = partial pressure of oxygen, PS = pulmonary stenosis, PVR = pulmonary vascular resistance.

Keywords: anesthesia, cesarean section, pregnancy, single ventricle
1. Introduction

Patients with an uncorrected single ventricle rarely survive to adulthood. Pregnancy and delivery rarely occur among these patients. Since single-ventricular pregnancy was first reported in 1963, only a few studies have addressed peripartum anesthesia management in these pregnant women.\textsuperscript{[1]} Systemic and pulmonary circulations originate from a single ventricle, where saturated and desaturated blood are mixed, leading to various degrees of systemic arterial desaturation.\textsuperscript{[2]} Pregnancy and its accompanying hemodynamic alterations increase the burden on this ventricle, resulting in a significantly increased risk of maternal morbidity or mortality.\textsuperscript{[3]}

2. Case report

The publication of this case was approved by the Ethics Committee of the Affiliated Hospital of North Sichuan Medical College, and written consent was signed by the patient for the purpose of research and publication.

A 35-year-old woman (height: 152 cm; body weight: 70 kg; New York Heart Association (NYHA) classification: II) was hospitalized at 37+6 weeks of pregnancy. She was diagnosed with congenital heart disease (CHD) at the age of 10 years and received no particular treatment. The patient was a gravida 5, para 0, abortion 1 female. She had given birth to three live children through vaginal delivery and experienced one spontaneous abortion. The physical examination revealed a heart rate (HR) of 90 beats/min, blood pressure (BP) of 130/84 mm Hg, respiratory rate of 21 breaths/min, oxygen saturation of 83%, and body temperature of 36.6°C. The arterial blood gas analysis (ABG) before oxygen therapy showed a pH of 7.46, partial pressure of carbon dioxide (PaCO\textsubscript{2}) of 28.0 mm Hg, and partial pressure of oxygen (PaO\textsubscript{2}) of 49.0 mm Hg. Her biochemical examination results, including routine blood tests, coagulation, hepatic and renal functions, serum electrolytes, myocardial damage markers, and brain natriuretic peptide, were normal. The patient reported no particular discomfort during the first and second trimesters, but she recently complained of reduced activity tolerance. As shown in Figure 1, echocardiography revealed a complex CHD. Specifically, there were findings of a single ventricle (the left ventricle is dominant), atrioventricular valve ectopic, double-inlet left ventricle, abnormal location of the great arteries, probably pulmonary stenosis (PS), atrial septal defect, and left-to-right shunt. A fetal ultrasound revealed a live intrauterine fetus with a breech presentation, who was small for gestational age and was wrapped by an umbilical cord around the neck for one circle. Based on the consensus of the multidisciplinary conference, the patient was scheduled for a cesarean section under regional anesthesia.

After entering the operating room, the patient received 100% oxygen via a facemask. Her vital signs before anesthesia included a HR of 105 beats/min, BP of 154/87 mm Hg, and oxygen saturation of 91%. Left radial arterial cannulation was then performed under local anesthesia to monitor BP dynamically. Prior to anesthesia, the ABG revealed a pH of 7.46, PaCO\textsubscript{2} of 26 mm Hg, PaO\textsubscript{2} of 55 mm Hg, arterial oxygen saturation of 88.7%, hemoglobin of 117 g/L, hematocrit of 38%, base excess in the extracellular fluid compartment of \(-5.3\) mmol/L, bicarbonate (HCO\textsubscript{3}\textsuperscript{−}) levels of 18.5 mmol/L, lactate levels of 1.7 mmol/L, potassium (K\textsuperscript{+}) levels of 3.6 mmol/L, and calcium (Ca\textsuperscript{2+}) levels of 1.11 mmol/L.

After hydration with 300 mL Ringer’s lactate, a combined spinal epidural anesthesia was performed in the left lying position with 2.0 mL of 0.67% ropivacaine, administered intrathecally at the L2-3 intervertebral space. This was followed by insertion of a 3-cm multiorifice epidural catheter. The patient was then placed in the supine position with a left tilt of 20°. Surgery began when the bilateral dermatomal-blockade level reached T4. Three minutes later, a 2.55-kg newborn girl was delivered. Once the umbilical cord was clamped, 100 μg of carbetocin was slowly injected to provide an adequate uterine tone. The maternal hemodynamics were generally stable, but she experienced a brief episode of postural hypotension (systolic BP reached as low as 85 mm Hg) upon changing her position from left tilt to supine prior to surgery. However, her hypotensive episode lasted less than 2 minutes because it was quickly corrected with intravenous ephedrine. The newborn maintained an APGAR score of 10 from 1 to 10 minutes after birth. The instant umbilical cord ABG revealed a pH of 7.37.

The surgical procedure lasted 80 minutes, and the total amounts of fluid input and output were 300 mL and 700 mL (400 mL of urine and 300 mL of estimated blood loss), respectively. At the end of the surgery, the ABG revealed a pH of 7.44, PaCO\textsubscript{2} of 25 mm Hg, PaO\textsubscript{2} of 69 mm Hg, arterial oxygen saturation of 95.4%, hemoglobin of 126 g/L, hematocrit of 39%, extracellular fluid compartment of \(-7.2\) mmol/L, HCO\textsubscript{3}\textsuperscript{−} of 17 mmol/L, lactate levels of 2.7 mmol/L, K\textsuperscript{+} of 3.7 mmol/L, and Ca\textsuperscript{2+} of 1.07 mmol/L. Postoperative pain relief was achieved by patient-controlled intravenous analgesia containing sufentanil and butorphanol. She and her baby were discharged from the hospital 5 days later. No congenital cardiac anomalies were detected in the baby.

3. Discussion and conclusions

A single ventricle or univentricular heart accounts for approximately 1% of CHD.\textsuperscript{[4]} Successful pregnancy and delivery are rare in uncorrected cases. A literature search was conducted in PubMed (https://www.ncbi.nlm.nih.gov/pubmed/) using the combination strategy with the following keywords: (pregnancy) AND ((single ventricle) OR (univentricular heart)) AND ((anesthesia) OR (cesarean section)). All available cases were summarized, and only 16 pregnant women with an uncorrected single ventricle underwent cesarean section, with 17 pregnancies (Table 1). They almost all are case reports.

![Figure 1. Parasternal long axis view of transthoracic echocardiography showing a single ventricle where the left ventricle is dominant. LA = left atrium, SV = single ventricle.](image-url)
| Author     | Anomaly                                                                 | Age  | Delivery time | NYHA | Exercise intolerance | Pulmonary hypertension | Cyanosis | Heart failure | $SPO_2$ | Anesthesia                                             | Outcome                  |
|------------|--------------------------------------------------------------------------|------|---------------|------|----------------------|------------------------|----------|---------------|---------|--------------------------------------------------------|--------------------------|
| Yuzpe[5]   | Single ventricle, laevo-TGA, PS, PDA, hypoplastic aortic arch             | 17   | G1P0, 40W     | NA   | Yes                  | No                     | Yes      | No            | 69%     | General: sodium pentothal, nitrous oxide               | M-good N-live, 2150g     |
| Leibbrandt[6] | Single ventricle, TGA, small PDA, mild aortic incompetence        | 25   | G2P0A1, 40W   | NA   | Probably             | No                     | No       | No            | ~84.5%  | Epidural: NA                                           | M-good N-live, 1540g     |
| Stiller[7]  | Single ventricle, subvalvar PS, TGA                                    | 29   | G4P1A2, 37W   | II–III| Yes                  | Yes                    | Yes      | Yes           | 84%     | Epidural: NA General anesthesia transferred from epidural anesthesia: NA | M-good N-live, 2353g     |
| Baumann[8]  | Single ventricle, TGA, VSD, mitral stenosis                           | NA   | NA, 38W       | II–III| Yes                  | Yes                    | Yes      | No            | 86%     | Epidural: NA                                           | M-good N-live, 2240g     |
| Tibaldi[9]  | Single ventricle, TGA, VSD, probably even Eisenmenger syndrome         | 31   | NA, 34W       | NA   | Probability          | Yes                    | Yes      | No            | NA      | General: NA                                            | M-good N-live, 820g      |
| Fong[10]    | Single ventricle, laevo-TGA, ASD, one AV valve, subvalvar PS           | 29   | G4P1A2, 36W   | IIa  | NA                   | No                     | Yes      | No            | 86%     | Epidural analgesia: bupivacaine Epidural anesthesia: 3% alkalized 2-chloroprocaine | M-good N-live, 1845g     |
| Peng[11]    | Single ventricle, moderate aortic stenosis, PDA                        | 24   | NA, 31W       | NA   | Yes                  | No                     | Yes      | No            | NA      | Epidural: NA                                           | M-good N-live, 934g      |
| Theodoridis[12] | Single ventricle, TGA, VSD                                             | 29   | G1P0, 38W     | NA   | Yes                  | No                     | Yes      | No            | 81-97%  | Epidural: NA Epidural: bupivacaine & sufentanil          | M-good N-live, 3070g     |
| Schummer[13] | Single ventricle, dextro-TGA, VSD                                     | 24   | G2P0, 32W     | II   | Probably             | Yes                    | Yes      | No            | 85%     | Epidural: NA Epidural: bupivacaine & sufentanil        | M-good N-live, 2035g     |
| Gomez[14]   | Single ventricle, TGA, PS, VSD                                        | 26   | G1P0, 27W     | II–III| Yes                  | No                     | Yes      | No            | 74-78%  | Epidural: NA                                           | M-good N-died at 48 h postpartum, 625g |
| Boukhina[15] | Single ventricle, Eisenmenger syndrome                                 | 27   | G1P0, 37W     | NA   | Yes                  | No                     | Yes      | No            | 70%     | Epidural: bupivacaine & sufentanil                      | M-good N-live, 3000g     |
| Wei[16]     | Single ventricle, single atrium, TGA, moderate PS                       | 20   | G1P0, 32W     | III  | Probably             | Yes                    | Yes      | No            | 80-90%  | Epidural: NA                                           | M-good N-died 2 days postpartum, 1345g |
| Wang[17]    | Single ventricle, severe pulmonary regurgitation, moderate MR and tricuspid regurgitation | 26   | G3P0A2, 34W    | II–III| Yes                  | No                     | Yes      | No            | 80%     | CSE: NA CSE: NA General: NA                            | M-good N-live, 1330g     |
|             | Single ventricle, tricuspid atresia, PS, ASD, PDA, mild MR              | 20   | G1P0, 34W     | III  | Yes                  | No                     | Yes      | No            | 86% (with oxygen) 82% | CSE: NA CSE: NA General: NA                            | M-good N-live, 1460 g    |
|             | Single ventricle, mitral atresia, ASD, severe PS                        | 34   | G2P1, 37W     | III–III| Yes                  | No                     | Yes      | No            | ~86%    | General: sodium pentothal, nitrous oxide               | M-good N-live, 1600 g    |
Based on the summary and analysis of the cases, the median maternal age was 26 (range: 17–35) years, and the median gestational age at delivery was 36 (range: 27–40) weeks. Apart from single ventricle, multiple congenital heart anomalies, such as transposition of the great arteries (10/17, 58.82%) and PS (8/17, 47.06%), were commonly detected. Seven patients had pulmonary hypertension (7/17, 41.18%), and three progressed to Eisenmenger syndrome (3/17, 17.65%). Most patients were cyanotic with a median basal oxygen saturation of 84.75% (range: 69%–97%). Except for four patients who received general anesthesia (4/18, 22.22%), all cases were conducted under regional anesthesia (14/18, 77.78%). There were two neonatal deaths (2/18, 11.11%), one died for pulmonary hemorrhage[14] and the other for a premature birth.[16] Although neuraxial block and epidural analgesia reportedly had favorable outcomes for single ventricular women undergoing vaginal delivery, the analgesia management for vaginal delivery is beyond the scope of this article.[10,19–21]

The natural history of patients with unoperated single ventricle is poor with a survival rate of ~30% in the first year of life and a median survival age of 14 years.[22] Several studies have shown that adult patients characterized with double-inlet left ventricular morphology and perfectly balanced circulation with some degree of PS could survive with mild-to-moderate left ventricular morphology and perfectly balanced circulation. Many adult patients characterized with double-inlet left ventricular morphology and perfectly balanced circulation have shown that adult patients characterized with double-inlet left ventricular morphology and perfectly balanced circulation. The anesthetic goal in patients with uncorrected single ventricle should be a balance between systemic vascular resistance and pulmonary vascular resistance (PVR) to avoid intracardiac shunt deterioration. Moreover, the cardiac output (CO) should be stabilized to avoid a forward flow reduction through either pulmonary or systemic circulation. Due to the potential of a right-to-left shunt, peripheral intravenous injection and infusion should minimize the risk of air embolism. Furthermore, appropriate postoperative analgesia contributes a lot to avoiding the catecholamine secretion that increases PVR, reduces pulmonary blood flow, and results in severe cyanosis.[26]

Since spinal anesthesia provides a reliable analgesic effect, and the epidural catheter is retained to maintain the anesthesia while extending the surgical time, the combined spinal epidural anesthesia method was chosen. Since regional anesthesia can cause systemic hypotension, fluid infusion was administered in advance, and invasive BP monitoring was performed before anesthesia. The dosage and volume of the intrathecal anesthetics were carefully titrated to maintain the dermatomal level below T4. The patient experienced supine hypotension syndrome after she was placed in the supine position. Then, a vasoconstrictor was immediately administered. The commonly used vasopressors for obstetrical patients include phenylephrine, methoxamine, ephedrine, and noxepinephrine. Carbetocin, an oxytocin analog, is routinely administered during cesarean section to reduce postpartum hemorrhage after delivering the fetus. Since oxytocin induces peripheral vasodilatation, hypotension, and increases CO (mediated by an increased HR and stroke volume), oxytocin agents should be infused under the slowest effective

| Author | Anomaly | Age | Delivery time | NYHA | Cyanosis | Pulmonary hypertension | Exercise intolerance | Heart failure | Cyanosis | Oxygen saturation | Anesthesia |
|--------|---------|-----|---------------|------|----------|-----------------------|---------------------|--------------|----------|------------------|------------|
| Minucci[18] | Single ventricle, Eisenmenger syndrome | 29 | GPPOL, 31W | NA | No | No | Yes | Yes | No | 70% | Spinal anesthesia: bupivacaine & fentanyl, i.v. | M-good N-live, 1640 g |
| Current case | Single ventricle, ASD, probably PS | 35 | GPPOL, 31W | II–III | Yes | No | Yes | No | No | 85% | CSE: bupivacaine | M-good N-live, 2550 g |

† Two pregnancies in one patient.

Fl ow accelerates in the 18 mm ID artery to its peak flow velocity of 3 m/s and a pressure gradient of 36 mm Hg.

4 symptomatology and preserved ventricular function.[22] with some degree of PS could survive with mild-to-moderate left ventricular morphology and perfectly balanced circulation have shown that adult patients characterized with double-inlet left ventricular morphology. The anesthetic goal in patients with uncorrected single ventricle should be a balance between systemic vascular resistance and pulmonary vascular resistance (PVR) to avoid intracardiac shunt deterioration. Moreover, the cardiac output (CO) should be stabilized to avoid a forward flow reduction through either pulmonary or systemic circulation. Due to the potential of a right-to-left shunt, peripheral intravenous injection and infusion should minimize the risk of air embolism. Furthermore, appropriate postoperative analgesia contributes a lot to avoiding the catecholamine secretion that increases PVR, reduces pulmonary blood flow, and results in severe cyanosis.[26]

Since spinal anesthesia provides a reliable analgesic effect, and the epidural catheter is retained to maintain the anesthesia while extending the surgical time, the combined spinal epidural anesthesia method was chosen. Since regional anesthesia can cause systemic hypotension, fluid infusion was administered in advance, and invasive BP monitoring was performed before anesthesia. The dosage and volume of the intrathecal anesthetics were carefully titrated to maintain the dermatomal level below T4. The patient experienced supine hypotension syndrome after she was placed in the supine position. Then, a vasoconstrictor was immediately administered. The commonly used vasopressors for obstetrical patients include phenylephrine, methoxamine, ephedrine, and noxepinephrine. Carbetocin, an oxytocin analog, is routinely administered during cesarean section to reduce postpartum hemorrhage after delivering the fetus. Since oxytocin induces peripheral vasodilatation, hypotension, and increases CO (mediated by an increased HR and stroke volume), oxytocin agents should be infused under the slowest effective
in single ventricular patients.\textsuperscript{[27,28]}
Ergometrine and prostaglandin F analogues (e.g., carboprost) should be avoided in patient with a single ventricle.\textsuperscript{[25]}
Besides, fluid balance is an important determinant of a favorable outcome.

In summary, this case showed that pregnant single ventricular women with a NYHA class of I–II could tolerate pregnancy and delivery. Both general and regional anesthesia are applicable to cesarean section in these patients. The principle of anesthesia management is to maintain the appropriate balance between systemic vascular resistance and PVR, and to maintain the preload and CO.

Author contributions
Conceptualization: Ji Wang.
Data curation: Yu Du, Yingzi Yang-Liu.
Methodology: Yu Du, Bin Chen.
Writing – original draft: Yu Du, Yingzi Yang-Liu.
Writing – review & editing: Bin Chen, Ji Wang.

References
1. Mandel A, Hirsch V. Cor triloculare biaatriatum Report of a case with survival to the age of 29 years. Am Heart J 1963;66:104–7.
2. Keepanasseril A, Raj A, Pillai AA, Baghel J, Plakkal N, Sartheesh S. Pregnancy outcome in women with an uncorrected single ventricle: a single-centre experience from South India. Obstet Med 2020;13:137–41.
3. European Society of Gynecology, Association for European Paediatric Cardiology, German Society for Gender Medicine, et al. ESC Guidelines on the management of cardiovascular diseases during pregnancy: the Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). Eur Heart J 2011;32:3147–97.
4. Steinberg EH, Dantzkler DR. Single ventricle with severe pulmonary hypertension: natural survival into the third decade of life. Am Heart J 1993;125:1451–3.
5. Yuzpe AA, Sanghvi VR, Johnson FL, Robinson JG. Successful pregnancy in a patient with single ventricle and other congenital cardiac anomalies. Can Med Assoc J 1970;103:1073–5.
6. Leibbrandt G, Munch U, Gander M. Two successful pregnancies in a patient with single ventricle and transposition of the great arteries. Int J Cardiol 1982;12:57–62.
7. Stuller RJ, Vorizilos AM, Nochimson DJ, Clement D, Campbell WA, Leach CN Jr. Single ventricle in pregnancy: case report and review of the literature. Obst Gynecol 1984;64:188–205.
8. Baumann H, Schneider H, Drack G, Alon E, Huch A. Pregnancy and delivery by caesarean section in a patient with transposition of the great arteries and single ventricle. Case report. Br J Obstet Gynaecol 1987;94:704–8.
9. Tihaldi G, Marchi L, Huscher M, Forlino G. [Anaesthesia for cesarean section in a pregnant woman with Eisenmenger’s syndrome. Description of a clinical case]. Minerva Ginecol 1988;40:145–6.
10. Fong J, Druzin M, Gimbel AA, Fisher J. Epidural anaesthesia for labour and caesarean section in a parturient with a single ventricle and transposition of the great arteries. Can J Anaesth 1990;37:680–4.
11. Peng TC, ChuaH EC, Tan PP. Epidural anesthesia for emergency caesarean section in a patient with single ventricle and aortic stenosis. Acta Anaesthesiol Sin 1997;35:39–44.
12. Theodordinis TD, Anagnostou E, Zeprimidis L, Dinas K, Bonits J. Successful pregnancy and caesarean section delivery in a patient with single ventricle and transposition of the great arteries. J Obstet Gynaecol 2005;25:69–70.
13. Schummer FW, Schummer C, Schleusner E, Frober R, Ferrari M, Fuchs J. [Uncorrected transposition of the great arteries and large ventricular septum defect peroperative management of a caesarean section]. Anaesthesist 2005;54:333–40.
14. Torres Gomez LG, Inigo Riesgo CA, Espinoza Ortegon MA, Barba Bustos AM. [Pregnancy in patients with single ventricle, corrected and uncorrected]. Ginecol Obstet Mex 2007;75:630–5.
15. Boukhris M, Hakim K, M’Sa’d H, Ouarda F, Boussaada R. Successful pregnancy and delivery in a woman with a single ventricle and Eisenmenger syndrome. J Saudi Heart Assoc 2013;25:261–4.
16. Wei C, Xue X, Zhu G, Li M, Liu H, Peng H. A primigravida with single atrium and single ventricle. Int J Gynaecol Obstet 2013;123:160–1.
17. Wang K, Luo H, Xin Y, Yu H. Successful pregnancy and delivery in patients with uncorrected single ventricle: three cases and literature review. Int J Cardiol 2015;184:135–9.
18. Minicucci S, Segala V, Verdecchia C, Simondon P, Casabona R, Sansone F. Safe management of cesarean section in a patient of Eisenmenger syndrome. Ann Card Anaesth 2012;15:296–8.
19. Copel JA, Harrison D, Whittemore R, Hobbins JC. Intrathecal morphine analgesia for vaginal delivery in a woman with a single ventricle. A case report. J Reprod Med 1986;31:274–6.
20. Dubois L, Belkaemhi H, Beli M, Dailland P, Carli P. [Single ventricle and obstetric anaesthesia: two cases report]. Ann Fr Anesth Reanim 2003;22:50–3.
21. Johnston TA, de Bono D. Single ventricle and pulmonary hypertension. A successful pregnancy. Case report. Br J Obstet Gynaecol 1989;96:731–4.
22. Gesuete V, Fabi M, Bonvicini M. A 74-year-old unoperated univentricular heart: the oldest reported survival. Cardiol Young 2016;26:805–7.
23. Poterucha JT, Anavekar NS, Egbe AC, et al. Survival and outcomes of patients with unoperated single ventricle. Heart 2016;102:216–22.
24. Ammash NM, Warnes CA. Survival into adulthood of patients with unoperated single ventricle. Am J Cardiol 1996;77:542–4.
25. Regitz-Zagrosek V, Roos-Hesselink JW, Bauersachs J, et al. 2018 ESC guidelines for the management of cardiovascular diseases during pregnancy. Kardiol Pol 2019;77:245–326.
26. Buckley R, Pickett JA. Pregnancy and the univentricular heart: case report and literature review. Int J Obstet Anesth 2000;9:55–63.
27. Heesen M, Carvalho B, Carvalho JCA, et al. International consensus statement on the use of uterotonics agents during caesarean section. Anaesthesia 2019;74:1305–19.
28. Joooste EH, Haft WA, Ames WA, Sherman FS, Vallejo MC. Anesthetic care of parturients with single ventricle physiology. J Clin Anesth 2013;25:417–23.