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Original Article

Anesthetic management for the peripartum care of women with Fontan physiology

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ABSTRACT

Background: As outcomes for surgical palliation have improved, women with single ventricle congenital heart disease are surviving into their reproductive years and may become pregnant. The cardiovascular changes of pregnancy may stress the Fontan circulation and pose significant risk to the mother and fetus.
Methods: Pregnant women with Fontan physiology were identified from the Ahmanson/UCLA Adult Congenital Heart Disease Center database. A total of 37 pregnancies were identified between 2000 and 2019. Twenty live births from 19 patients were reviewed and compared for cardiac history, obstetric history, anesthetic management and cardiovascular outcomes.
Results: Median gestational age at delivery was 35 weeks. Ten of 20 births were by cesarean delivery. An epidural technique was used as the primary anesthetic for 19 deliveries and general anesthesia was used for one cesarean delivery. An arterial line was placed in the peripartum period for three deliveries. Central venous access was established in the peripartum period for one patient. The mean blood loss for cesarean deliveries was 626 mL (range 240–1200 mL). The mean net peri-operative intake/output was positive 93.5 mL. Three patients were briefly transferred to the intensive care unit postpartum for higher level monitoring and care.
Conclusion: Epidural anesthesia is safe and effective for both vaginal and cesarean deliveries. Judicious fluid management is critical in minimizing postpartum cardiovascular complications. Many patients do not require a higher level of care, invasive monitoring or central venous access during the peripartum period.

Introduction

Single ventricle physiology is present in congenital heart defects such as hypoplastic left heart, tricuspid atresia, pulmonary atresia, double outlet right ventricle, double inlet left ventricle, severe Ebstein's anomaly, and sometimes complete atrioventricular canal defects. In 1971, Francis Fontan and Eugene Baudet devised a procedure to divert systemic venous return directly into the pulmonary artery; a passive conduit directs systemic venous return to the pulmonary circulation, allowing the remaining ventricle to drive systemic circulation.[1–4] Without a ventricular pump, pulmonary blood flow depends on the transpulmonary gradient, pulmonary vascular resistance (PVR), and the generation of negative intrathoracic pressure during spontaneous inspiration.[5,6] Consequently, the Fontan circulation represents a chronic low cardiac output state with limited reserve.[1]

Fontan palliation has two major forms, the right atrial to pulmonary artery (RA-PA) connection described by Fontan and total cavo-pulmonary connection (TCPC). The RA-PA Fontan palliation and its modifications consist of a valved or non-valved right atrial to pulmonary artery connection. However, surgical atriotomy and inclusion of the atrium in the Fontan circuit results in progressive right atrial dilation, fibrosis, and atrial arrhythmias. This led to the development of TCPC which excludes much or all of the right atrium from the Fontan circuit. The two variations of TCPC, namely lateral tunnel and extracardiac, are characterized by a superior vena cava to pulmonary artery anastomosis and an inferior vena cava to pulmonary artery conduit that reduce complications related to atrial inclusion (Fig. 1).[6]

The TCPC has led to improved long-term outcomes. Actuarial 20-year post-Fontan survival rates are estimated at 82.6% in early survivors of the Fontan operation.[7] Increasingly, women with Fontan circulation are reaching their reproductive years and considering pregnancy. Studies have shown higher rates of spontaneous abortion (15–25%) and intra-uterine fetal demise in patients with congenital heart disease.[8] However, studies consistently report that clinically

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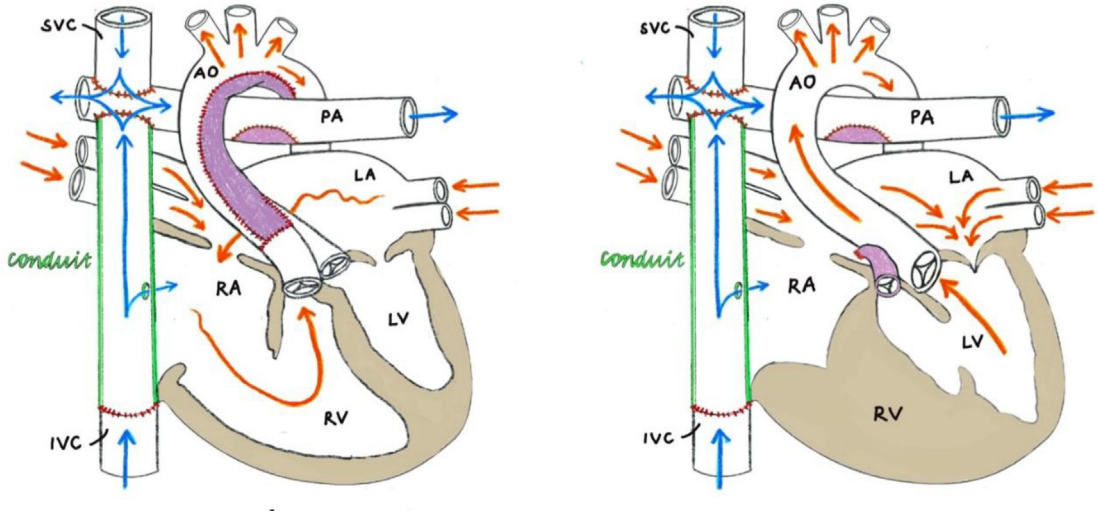

Fig. 1. A) Fontan Palliation Hypoplastic Right Heart Disease B) Fontan Palliation Hypoplastic Left Heart Disease. Total cavo-pulmonary connection is shown. The blue arrows represent de-oxygenated blood flow returning to the heart and passing through the pulmonary circulation. The red arrows represent oxygenated blood flow through the systemic circulation. Pregnancy is associated with increased blood volume and cardiac output; in women with Fontan physiology, these demands must be accommodated without the support of a subpulmonic ventricle. LV: left ventricle; RV: right ventricle; LA: left atrium; RA: right atrium; AO: aorta; PA: pulmonary artery; SVC: superior vena cava; IVC: inferior vena cava. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

stable patients with good functional capacity at the time of conception have favorable pregnancy outcomes.[9] The normal physiologic changes of pregnancy include alterations in maternal volume status, vascular resistance, and cardiac output, all of which must be considered within the framework of the Fontan circulation (Table 1). Despite the growing body of literature, anesthetic management has not been well described and the standard care for clinically stable patients has not been defined. Most recently, Tiouririne et al. published a review of 27 case reports describing current peripartum management practices for patients with Fontan at multiple institutions; details regarding obstetric history, cardiac history, and anesthetic management were not specified for all cases.[10]

Also, Monteiro et al. published a detailed single-center case series of eight patients with 14 deliveries.[11] This retrospective study aimed to provide a more comprehensive overview of the peripartum management of Fontan patients. Our study highlights differences in antepartum management, especially regarding anticoagulation and obstetric management resulting in small differences in anesthetic management, however it also reinforces several important concepts underscored in the Monteiro et al. case series.

Methods

Institutional review board approval was obtained and written informed consent requirement was waived. Women with single ventricle physiology who had undergone a Fontan palliation and reported a pregnancy were identified from the Ahmanson/UCLA Adult Congenital Heart Disease (ACHD) pregnancy database and included in this review. A total of 37 pregnancies were identified between 2000 and 2019. Twenty-four of 37 (67%) pregnancies resulted in live births, 10/37 (28%) pregnancies resulted in spontaneous abortions, 2/37 (5%) pregnancies underwent elective termination, and 1/37 (3%) resulted in intra-uterine fetal demise. Among the 24 live births, four deliveries were managed at outside institutions and were excluded, leaving 19 patients (20 live births) in the final analysis.

Medical record review encompassed the underlying cardiac defect, type of surgical palliation, pre-pregnancy oxygen saturation, functional status, and obstetric and cardiac complications in the intra-, peri- and postpartum periods. Delivery data included anesthetic techniques, utilization of invasive lines, blood loss, fluid administration, and medication administration.

Results

Antepartum

In this series, 11/19 (58%) patients had a lateral tunnel Fontan, 5/19 (26%) had an extracardiac Fontan and 3/19 (16%) had a RA-PA Fontan. Preserved single systolic ventricular function with a NYHA Classification of 1 was seen in 16/19 (84%) patients, while 3/19 (16%) were classified as NYHA Class 2 (Table 2). Thirteen patients were on low-dose aspirin, eight were on a beta-blocker (Table 3). No patients were on prophylactic or therapeutic anticoagulation.

Pregnancy termination was recommended to patient #9 due to poor pre-pregnancy functional status and poorly controlled atrial fibrillation, but she declined. Patient #9 was on flecainide 100 mg twice daily prior to conception and her dose increased to 200 mg twice daily during the antepartum period after two recurrent episodes of atrial flutter, both of which required cardioversion. The antepartum periods of the other patients were uneventful.

Intrapartum

The median gestational age at delivery was 35 weeks (range: 27 weeks 4 days to 40 weeks 1 day) (Table 4). Ten of 20 (50%) pregnancies were delivered pre-term. Pre-term deliveries were due primarily to pre-term premature rupture of the membranes (50%) followed by non-reassuring fetal heart tracing (20%). Patient #4′s delivery was complicated by preeclampsia with severe features.

Neuraxial anesthesia was used in 19 deliveries without complications. General anesthesia was used in one cesarean delivery (Table 5). Epidural analgesia was initiated for vaginal deliveries with 0.0625% bupivacaine with 2 μg/mL fentanyl given in fractionated doses, exceeding a total volume of 10 mL, to achieve the desired clinical effect. A test dose was not given to avoid the administration of epinephrine. For the nine cesarean deliveries, plain lidocaine 2% was

Table 1
Common organ dysfunction and possible comorbidities of the Fontan circulation

Organ	Fontan physiology	Antepartum Fontan physiology	Peripartum Fontan physiology
Brain	* Developmental delay (multiple CPB runs, chronic hypoxemia, CPB associated micro-emboli) * Congenital heart disease associated CNS deficits (congenital brain–heart disease) * CVA secondary to thrombotic events/vascular shunts	* Exposure to early adversity and physiological risk may affect long-term responses to stress * CVA	* Mental well-being in clinical environment (ICU/labor floor) * Psychosocial stress related to complex pregnancy
Heart	* Absence of a subpulmonic ventricle * Obligatory venous hypertension (CVP 10–15 mmHg) * Preload deprivation of the univentricular heart * Decreased cardiac output at rest and exercise * Potential right to left shunt (baffle fenestration)	* Arrhythmias (neuro-hormonal mechanism) * Presence and/or potential to develop or exacerbate low systemic cardiac output * In left hypoplastic left heart disease, presence of diastolic dysfunction and high systemic vascular resistance	* Progressive heart failure (third trimester and postpartum) * Supraventricular arrhythmia * Bradycardia * Longer-term effect of pregnancy on Fontan physiology
Lungs	* Non-pulsatile flow to the lungs * Restrictive or obstructive lung physiology * Possible diaphragmatic paralysis * Progressive cyanosis due to veno-venous and pulmonary arterial malformations * Plastic bronchitis * Resistance of pulmonary flow is relatively fixed i.e. increased cardiac output causes pulmonary resistance and CVP to rise * Chylothorax * Increased risk for pulmonary embolism		* Severe decrease in functional residual capacity * Peripartum pulmonary embolus
Liver	* Hepatic congestion * Chronic liver disease secondary to intrahepatic thrombosis * Higher prevalence of hepatitis C * Hepatotoxicity of anti-arrhythmic drugs (e.g. amiodarone)		
Kidney	* Proteinuria * Fixed reduced cardiac output causing decrease in glomerular filtration rate		
Hematology	* Anemia * Thrombophilic tendency * Bleeding propensity	* Thrombo-embolism	* Postpartum hemorrhage (concomitant anticoagulation and/or antiplatelet therapy)
Bowel	* Protein-losing enteropathy		
Vascular	* Venous hypertension causing varicosities and ulcers * Decreased lymphatic return * Hypertension		* Deep venous thrombosis * Pregnancy-induced hypertension
Endocrine	* Diabetes * Hyperlipidemia * Obesity	* Gestational diabetes	*Gestational diabetes
Fetus	* Late menarche * Primary amenorrhea * Infertility	* Fetal loss * Fetal complications	* Fetal growth restriction possibly related to placental insufficiency * Prematurity

CPB: cardiopulmonary bypass; CVA: cerebrovascular accident; ICU: intensive care unit; CVP: central venous pressure.

Table 2
Baseline cardiac characteristics of study population

Patient	Age	Primary etiology	Surgical palliation	Baseline SpO_2	NYHA class	Ejection fraction
1	24	Double outlet RV	Lateral tunnel Fontan	89	2	30–35%
2	27	Pulmonic and tricuspid atresia	Extracardiac Fontan	92	1	55–60%
3	20	Double outlet RV and pulmonic atresia	Extracardiac Fontan	95	1	50–55%
4	20	Pulmonic and tricuspid atresia	Lateral tunnel Fontan	93	1	60–65%
5	26	Pulmonic atresia	RA-PA Fontan	92	2	35–40%
6	31	Double inlet LV and pulmonic atresia	Lateral tunnel Fontan	95	1	50–55%
7	27	Pulmonic atresia	Lateral Tunnel Fontan	91	1	50–55%
8	18	Hypoplastic RV	Extracardiac Fontan	91	1	45–50%
9	20	Tricuspid atresia	RA-PA Fontan	94	1	55–60%
10	22	Tricuspid atresia	Extracardiac Fontan	97	1	30–35%
11	23	Pulmonic atresia	Lateral Tunnel Fontan	94	1	55–60%
12	29	Double inlet LV	Lateral Tunnel Fontan	95	1	50–55%
13	27	Tricuspid atresia	RA-PA Fontan	92	1	55–60%
14	26	Pulmonic atresia	Lateral tunnel Fontan	93	1	60–65%
15	22	Pulmonic atresia	Lateral tunnel Fontan	90	1	50–55%
16	18	Unbalanced AV canal and hypoplastic RV	Lateral tunnel Fontan	92	1	55–60%
17	24	Tricuspid atresia	Lateral tunnel Fontan	94	1	55–60%
18	15	Double inlet LV and aortic coarctation	Lateral tunnel Fontan	89	2	50–55%
19	30	Double inlet LV and pulmonic atresia	Extracardiac Fontan	97	1	55–60%

SpO_2: oxyhemoglobin saturation; NYHA: New York Heart Association; Age: maternal age at delivery; RV: right ventricle; LV: left ventricle.

Table 3
Additional cardiac history of study population

Patient	Anticoagulation	Additional medications	Baseline ECG
1	None	Digoxin 0.25 mg QD, Carvedilol 3.125 mg BID	Sinus, NSTW
2	Aspirin 81 mg	Carvedilol 25 mg BID	Sinus, prolonged QT
3	Aspirin 81 mg	None	Sinus
4	Aspirin 81 mg	None	Sinus, T-wave inversions
5	Aspirin 81 mg	Metoprolol succinate 12.5 mg QD	Sinus, RBBB
6	Aspirin 81 mg	Metoprolol succinate 25 mg BID	A paced
7	Aspirin 81 mg	Metoprolol succinate 25 mg QD, Digoxin 0.125 mg QD	Atrial ectopic
8	None	None	Sinus
9	None	Flecainide 200 mg BID, Digoxin 0.125 mg QD	AV Sequential Paced
10	Aspirin 81 mg	Digoxin 0.125 mg QD	Sinus, left axis deviation
11	Aspirin 81 mg	Metoprolol tartate 12.5 mg BID	Sinus, left axis deviation
12	None	None	Sinus
13	Aspirin 81 mg	None	Sinus
14	Aspirin 81 mg	None	Sinus
15	None	None	Sinus
16	Aspirin 81 mg	None	Sinus
17	None	Labetelol 50 mg PRN	Sinus, NSTW
18	Aspirin 81 mg	None	AV Sequential Paced
19	Aspirin 81 mg	Labetelol 200 mg BID	Sinus, NSTW

ECG: electrocardiogram; QD: daily; BID: twice daily; NSTW: non-specific T-wave abnormalities; RBBB: right bundle branch block; AV: atrioventricular.

used to achieve surgical anesthesia, as is the standard practice for laboring patients who require cesarean deliveries at our institution. The volume of lidocaine ranged from 15 to 25 mL (Table 5). Epidural morphine was also routinely given for postoperative pain control. The mean blood loss at cesarean deliveries was 626 mL (range 240–1200 mL) (Table 6). The mean net peri-operative intake/output was positive 93.5 mL. Inotropic and vasopressor support was administered for only one delivery (patient #10).

Patient #9 lost 1200 mL of blood from a perineal laceration at delivery and developed hemolysis, elevated liver enzymes and low platelet count (HELLP) syndrome. Symptomatic bradycardia developed due to supratherapeutic magnesium levels and loss of pacemaker capture. She was emergently intubated for transcutaneous pacing and transferred to the intensive care unit. After urgent dialysis pacemaker capture was restored.

Patient #10 was admitted with a segmental pulmonary embolism and increasing oxygen requirements. She required 6 L of oxygen via facemask to maintain her baseline oxygen saturation of 95%. Urgent delivery was warranted because respiratory failure posed a risk to her and her fetus; any resulting PVR elevation was likely to be poorly tolerated due to her Fontan circulation. Patient #10′s cesarean delivery was performed under general anesthesia, with arterial and central lines for monitoring. General anesthesia was induced with etomidate and vecuronium and she received a dopamine infusion for the duration of the cesarean section. At the end of surgery, she remained intubated and was transferred to the intensive care unit for recovery.

An arterial line was placed for two additional cesarean deliveries (patients #1 and #13). Both arterial lines were removed prior to leaving the operating room. Patient #1 was also transferred to the intensive care unit for precautionary monitoring.

Postpartum

Patients #14, #16 and #18 developed symptoms of volume-overload for which intravenous (IV) furosemide was administered. Patients #14 and #16 received a single dose of IV furosemide 20 mg. Patient #18 received two doses of IV furosemide 20 mg. Patients #4, #14, #16 and #18 required supplemental oxygen to maintain baseline oxygen saturation.

Patient #9 was extubated on postpartum day one and transferred to the postpartum floor on postpartum day two. The remainder of her postpartum course was uneventful. Patient #10 was extubated on postpartum day one and started on therapeutic anticoagulation. The remainder of the postpartum course was uneventful, and she was discharged 10 days after delivery.

The median delivery to discharge time for all deliveries was four days (range 1–10 days). The median delivery to discharge interval for patients who delivered vaginally was two days (range 1–7 days), while for those delivered by cesarean the median discharge time was three days (range 3–10 days).

Table 4
Obstetric history of study population

Patient	Mode of delivery	Gestation at delivery (weeks and days)	Indication for early delivery	Indication for cesarean
1	CCS	27 and 4	PPROM	Breech
2	LTCS	34 and 2	Non-reassuring fetal heart tracing	Choice
3	LTCS	40 and 1	-	Breech
4	VV	37 and 5	Preeclampsia with severe features	-
5	VF	32 and 2	PPROM	-
6	VF	35 and 3	PPROM	-
6	LTCS	34 and 0	PPROM	Breech
7	LTCS	34 and 5	IUGR and elevated Dopplers	Choice
8	VF	37 and 5	Non-reassuring fetal heart tracing	-
9	VF	37 and 4	HELLP Syndrome	-
10	CCS	30 and 0	Newly diagnosed PE	New supplemental O_2 requirement
11	LTCS	38 and 6	-	History of myomectomy
12	V	38 and 0	-	-
13	LTCS	33 and 5	Absent end diastolic flow	Breech
14	VF	38 and 1	-	-
15	V	38 and 3	-	-
16	LTCS	37 and 5	-	Elective repeat
17	LTCS	35 and 6	Non-reassuring fetal heart tracing	Breech
18	V	36 and 0	PPROM	-
19	VV	37 and 0	-	-

V: vaginal unassisted; VF: vaginal forceps assisted; VV: vaginal vacuum assisted; LTCS: low transverse cesarean section; CCS: classical cesarean section; IUGR: intrauterine growth restriction; PPROM: preterm premature rupture of membranes; PE: pulmonary embolism; HELLP: hemolysis, elevated liver enzymes, low platelets; O_2: oxygen.

Table 5
Anesthetic management for vaginal and cesarean deliveries

Patient	Mode of delivery	Analgesia or anesthesia	Invasive vascular lines	EBL
4	VV	Bupivacaine 0.0625%, fent 2 μg/mL (8 mL)	None	200
5	VF	Bupivacaine 0.25 % (9 mL) Second Stage: Bupivacaine 0.125 % (6 mL)	None	200
6	VF	Bupivacaine 0.0625%, fent 2 μg/mL (9 mL) Second Stage: Lidocaine 1% (10 mL)	None	800
8	VF	Bupivacaine 0.0625%, fent 2 μg/mL (8 mL)	None	500
9	VF	Bupivacaine 0.0625 %, fent 2 μg/mL (5 mL)	None	1200
12	V	Bupivacaine 0.0625%, fent 2 μg/mL (10 mL) Second Stage: Bupivacaine 0.125% (10 mL)	None	400
14	VF	Bupivacaine 0.0625%, fent 2 μg/mL (5 mL) Second Stage: Bupivacaine 0.125% (10 mL)	None	450
15	V	Bupivacaine 0.0625%, fent 2 μg/mL (10 mL)	None	700
18	VF	Bupivacaine 0.0625%, fent 2 μg/mL (10 mL) Second Stage: Bupivacaine 0.25% (10 mL)	None	450
19	V	Bupivacaine 0.0625%, fent 2 μg/mL (10 mL)	None	300
1	CCS	Epidural 2% lidocaine 20 mL	Arterial line	400
2	LTCS	Epidural 2% lidocaine 25 mL	None	800
3	LTCS	Epidural 2% lidocaine 20 mL	None	325
6	LTCS	Epidural 2% lidocaine 23 mL	None	750
7	LTCS	Epidural 2% lidocaine 15 mL	None	550
10	CCS	General (40 mg etomidate, 7 mg vecuronium)	Arterial line and central venous access	1200
11	LTCS	Epidural 2% lidocaine 17 mL	None	550
13	LTCS	Epidural 2% lidocaine 25 mL	Arterial line	240
16	LTCS	Epidural 2% lidocaine 25 mL	None	675
17	LTCS	Epidural 2% lidocaine 20 mL	None	770

Patient #10 is the only patient who required pressors during the peripartum period. Patient was on dopamine drip for the duration of the cesarean section.
V: vaginal unassisted; VF: vaginal forceps assisted; VV: vaginal vacuum assisted; LTCS: low transverse cesarean section; CCS: classical cesarean section; fent: fentanyl; EBL: estimated blood loss in mL.

Table 6
Fluid management for cesarean deliveries

Patient	Crystalloid (mL)	5% albumin (mL)	EBL (mL)	Urine output (mL)	Net intake/output (mL)
1	1200	-	400	200	Positive 600
2	1100	-	800	400	Negative 100
3	750	-	325	125	Positive 250
6	1300	-	750	180	Positive 370
7	500	-	550	100	Negative 150
10	1000	250	1200	50	Even
11	1000	-	550	50	Positive 400
13	500	-	240	300	Negative 40
16	750	-	675	250	Negative 175
17	800	-	770	250	Negative 220

EBL: estimated blood loss. Mean blood loss for all cases was 626 mL. The mean net fluid intake/output balance in mL for all cases was positive 93.5 mL.

Discussion

Management of parturients with Fontan circulation must balance maternal cardiac work with the demands for placental perfusion. The Fontan procedure is palliative rather than curative and cardiopulmonary adaptation is highly variable. Pre-pregnancy single ventricular systolic function, as assessed by ejection fraction and pulmonary pressures, may predict a patient's ability to tolerate the physiologic changes associated with pregnancy and delivery.[8,9] At our institution, we counsel patients based on a detailed history of arrhythmias, clinical symptoms of ventricular dysfunction, and comprehensive stress testing (ideally prior to conception). These early assessments help inform patients desiring conception or debating continuation of pregnancy. Medication adjustments may occur before or after pregnancy is confirmed. Anticoagulation is controversial due to antepartum and postpartum bleeding risks and currently there is no consensus on routine anticoagulation for the Fontan population. The most recent American guidelines on pregnancy and complex congenital heart disease indicate that a history of thrombo-embolic events justifies continuation of anticoagulation.[8] Known teratogenic medications such as angiotensin-converting enzyme inhibitors or angiotensin receptor blockers are discontinued. Transthoracic echocardiograms are performed once every trimester and at admission to monitor changes in cardiac function. The aim is to reduce peripartum cardiovascular complications by means of careful patient selection, pre-pregnancy cardiovascular optimization, and individualized labor and delivery plans that usually include vaginal delivery and epidural analgesia.

The normal physiologic changes of pregnancy stress the Fontan circulation and increase the risk of heart failure and arrhythmia. Higher blood volume may raise pressure within the Fontan circuit, increasing atrial wall stress and, consequently, increasing the risk of atrial arrhythmias. Labor and delivery present additional demands from Valsalva strain, pushing, and breath-holding which impairs venous return and elevates PVR. This restricts blood flow into the pulmonary circulation. During Valsalva and breath-holding, the negative intrathoracic pressure that normally propels Fontan flow is lost, reducing pulmonary blood flow and lowering the fraction of oxygenated blood returned to the systemic ventricle for cardiac output. Nonetheless, vaginal delivery is preferred over cesarean delivery to minimize expected blood loss and to decrease the need for higher doses of neuraxial anesthesia

which often cause significant hemodynamic effects. Assistance with the second stage of labor is recommended for vaginal delivery to avoid Valsalva strain, pushing, and breath-holding. An alternative to forceps or vacuum assisted delivery is Valsalva with vocalization; this allows the glottis to remain open during Valsalva, minimizing impairment to venous return. Analgesia is also an important consideration during labor and delivery. Pharmacologic augmentation of uterine tone with oxytocin infusion, without boluses, is preferred. Second-line agents like methylergonovine and carboprost tromethamine may be necessary for peripartum hemorrhage, however it is important to be cognizant of the associated hemodynamic effects which can compromise pulmonary blood flow and cardiac output.

Analgesia and anesthesia

For planned vaginal deliveries, we advocate early epidural catheter placement, omission of an epinephrine-containing test dose, and incremental titration of local anesthetics to verify catheter function and to avoid sudden cardiovascular problems. For cesarean deliveries general or neuraxial anesthesia may be used, however suboptimal oxygenation and ventilation may occur during airway management. Hypoxia and hypercarbia increase PVR, impair Fontan flow, and decrease cardiac output. Positive pressure ventilation can also impair passive flow through the Fontan. Employing neuraxial anesthesia avoids airway manipulation and provides analgesia to help offset sympathetic stimulation, which can increase systemic and pulmonary vascular resistance.[10,11] We recommend gradual epidural anesthesia over spinal anesthesia to prevent a sudden decrease in systemic vascular resistance resulting in decreased preload. While we acknowledge that spinal anesthesia is more desirable for cesarean section, patients with Fontan physiology may tolerate these rapid changes poorly (spinal anesthesia may be appropriate with right hypoplastic heart disease, when the natural left ventricle is pumping against systemic circulation.) Pre-emptive hydration with 250–500 mL IV fluid can offset the decrease in systemic vascular resistance from sympathetic blockade, but fluids must be used judiciously to avoid volume overload. Phenylephrine may be used to address hypotension and augment venous return, however the effect on PVR is concerning in this patient population. Fractionated epidural dosing may preclude the need for vasopressor support and allow non-invasive blood pressure monitoring to suffice.

Need for invasive monitoring

Although complex, Fontan physiology does not inherently demand invasive monitoring commonly reported in older case series. Invasive line placement carries its own risks, and there is increasing evidence that many patients have good outcomes without the use of invasive monitors and/or central venous access.[10,11] Our institutional experience also supports selective use of invasive monitoring. Central venous pressure reflects pressure in the Fontan circulation and may vary tremendously with pain, breath-holding, and Valsalva strain, so central venous pressure is an especially poor indicator of volume status. Routine placement of pulmonary arterial catheters is not recommended due to the significant anatomic modifications in the Fontan circulation and the potential for thrombo-embolic complications. The need for invasive arterial blood pressure monitoring and/or central venous access should be assessed for each patient individually. An arterial line was placed for patient #1 out of an abundance of caution, considering it was our institution's first experience with managing a parturient with Fontan physiology. In recent years we have moved away from invasive monitoring based on periodic review of our practices and outcomes. The vast majority of these patients managed with epidural analgesia using incremental epidural titration have not required invasive hemodynamic monitoring and have had good outcomes. A thorough history and physical exam are critical when evaluating peripartum patients with Fontan physiology. Brain natriuretic peptide levels greater than 128 pg/mL are a predictor of maternal cardiovascular events, which we have been tracking in more recent cases, and should also be considered in the evaluation.[12] Based on this case series, the benefit-to-risk ratio favors reserving invasive monitoring for patients who are at high risk of cardiovascular complications that might necessitate vasopressor or inotropic support. Only patient #10 required vasopressor support during the peripartum period and only required it while under general anesthesia. Although dopamine was used in this case, the consensus among the obstetric anesthesia group at our institution is that norepinephrine is the preferred first-line vasopressor for undifferentiated hypotension. In future cases we will consider the use of non-invasive continuous blood pressure monitoring devices that are now available, especially in situations where vasopressor support becomes increasingly necessary.

Need for higher-level care

Atrial arrhythmias and symptomatic heart failure are the most common cardiac complications but there were no peripartum deaths in any of the case series reviewed or the present cohort.[13,14] In one series, 10% of women experienced cardiovascular complications, most commonly an atrial arrhythmia.[13] History of atrial arrhythmias was the best predictor of peripartum atrial arrhythmias, with most occurring in women with RA-PA connections.[13,14] In this series, patients #9 and #10 were transferred to the intensive care unit for respiratory support. Most patients did not require a higher level of care in the postpartum period, and were safely managed on a regular postpartum floor with telemetry.

Limitations are inherent in a case series. There is no control population. The study is subject to selection bias. There is a possibility of inaccuracies in documentation prior to 2013 because documentation was done on paper charts. Patients included in the study were often managed concurrently by both the Maternal-Fetal Medicine and Adult Congenital Heart Disease teams during the antepartum period. Patients were followed carefully and optimized prior to admission to the labor and delivery floor. Our management, approach and outcomes may not be applicable for patients admitted to outside institutions with limited workup and without prior optimization. The disproportionate number of patients with tricuspid atresia may also limit generalizability.

The physiologic challenges that pregnancy presents to women with Fontan circulation warrant continued exploration because further knowledge is imperative to improving outcomes. The next phase of investigation is a prospective comparison of pre-pregnancy optimization and peripartum management strategies in patients with varying pre-pregnancy functional capacities. Examination of the utility of point-of-care transthoracic echocardiogram may prove useful. Using ultrasound to assess for the presence of diffuse B-lines or distension of the inferior vena cava may help guide care of this high-risk population. Use of non-invasive continuous blood pressure measurement might be beneficial for closer monitoring during delivery.

Only women with well-compensated Fontan physiology are advised to consider pregnancy. Fontan patients who pursue pregnancy assume significant risk and the patient's priorities may change if complications arise. It is prudent to continually reassess management goals as the clinical condition of the mother and fetus evolve. Complex care plans require a multidisciplinary team with clinical acumen for Fontan physiology; accordingly, our team includes a nurse specialist, adult congenital cardiologists, maternal-fetal medicine obstetricians, intensivists, and obstetric anesthesiologists. Even with well-compensated physiology and preserved ventricular function, this population remains at risk for peripartum heart failure, arrhythmias, and thrombo-embolism. Epidural analgesia and assisted vaginal delivery and epidural anesthesia produce a desirable balance between maternal systemic and pulmonary vascular resistances. Judicious fluid management is also important in minimizing postpartum complications. Invasive hemody-

namic monitoring and central vascular access should be considered on a case-by-case basis. Based on our experience, we endorse invasive monitoring when ejection fraction is significantly reduced from baseline or there is evidence of pulmonary edema. We also consider invasive monitoring for patients at high-risk for postpartum hemorrhage or with concomitant pathologies such as preeclampsia with severe features, HELLP syndrome, and abnormal placentation. We would likely employ continuous blood pressure monitoring if we planned spinal anesthesia with a corresponding prophylactic vasopressor infusion. In our institution, patients with invasive monitoring lines need to recover in a different nursing unit, possibly under the care of intensivists whose invasive monitoring preferences may influence the decision to place them pre-operatively. However, most patients can recover on a regular postpartum floor unless the patient is deemed high risk for arrhythmia, thrombo-embolism, or decompensated heart failure.

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Declaration of interests

None.

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