Oxford University Hospitals NHS
NHS Foundation Trust

Acott C, Catton H, Chachlani M*, Turnbull C

Fontan Circulation in Caesarean Section: A Physiology Driven Approach

BACKGROUND

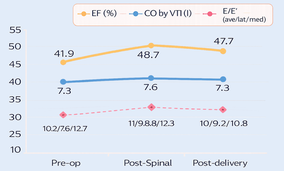
The Fontan circulation is characterised by passive pulmonary blood flow, with cardiac output dependent on preload, low PVR, ventricular function, and SVR. Pregnancy may destabilise this circulation, increasing the risk of hypoxia, arrhythmia, thromboembolism, cardiac decompensation, and foetal growth restriction. Caesarean section under general or regional anaesthesia adds further haemodynamic challenge.

CASE 1

A 31-year-old nulliparous woman with pulmonary atresia (lateral tunnel Fontan) developed hypoxia and severe IUGR, requiring delivery at 34 weeks.

Intraoperative TTE showed a dilated single ventricle (EF 40–45%). A combined spinal–epidural (CSE) with repeat spinal anaesthesia was used.

Ventricular contractility improved modestly post-spinal, with minimal haemodynamic change after delivery.



CASE 2

An 18-year-old nulliparous woman with pulmonary atresia, VSD and extracardiac fenestrated Fontan developed hypoxia and worsening IUGR, requiring delivery at 28^{+6} weeks.

A low-dose CSE anaesthesia was used, with a vasopressin infusion providing intraoperative stability.

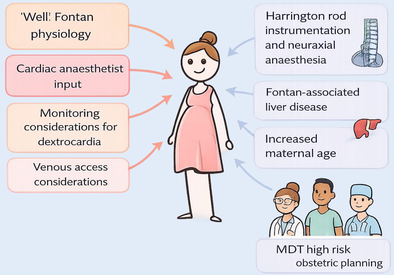
Maternal and neonatal outcomes were favourable following an uneventful delivery.

CASE 3

A 34-year-old primiparous woman with complex congenital heart disease (dextrocardia, TGA, Fontan), with Fontan-associated liver disease and kyphoscoliosis, underwent elective Caesarean section.

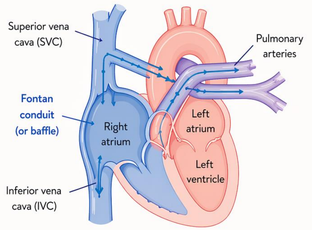
A CSE provided anaesthesia, with invasive arterial monitoring. ECG monitoring was adapted for dextrocardia.

The procedure was stable, and recovery was uncomplicated aside from transient atrial ectopy.



The Fontan Circulation

Surgical connection that directs systemic venous blood to the pulmonary arteries, bypassing the right ventricle.



Systemic venous return → Pulmonary arteries (passive flow, no pump) → Left heart → Systemic circulation

Determinants of Fontan Circulation

Systemic Venous Pressure
- Higher pressures impede pulmonary flow
- Aim to keep as low as possible

Pulmonary Vascular Resistance
- Must be low and non-reactive
- Avoid hypoxia, acidosis, hypercarbia

Blood Volume / Preload
- Adequate volume needed for optimal output
- Both hypovolemia and fluid overload harmful

Systemic Ventricular Function
- Poor ventricular function → low output
- Maintain sinus rhythm and contractility

Atrioventricular Valve Function
- Regurgitation reduces forward output
- Maintain optimal valve function

CONCLUSION

Caesarean section in Fontan physiology can be safely undertaken with careful planning and a physiology-driven approach. Patient counselling and preparation are essential within a shared decision-making model. Maintenance of preload, avoidance of increased pulmonary vascular resistance, and preservation of systemic vascular resistance are key.

Anaesthetic management should be individualised and physiology-guided.

Postpartum follow-up, including cardiology review and counselling regarding future pregnancy, is essential.

Fontan-Associated Complications

Fontan-Associated Liver Disease (FALD)
Chronic venous congestion → fibrosis, cirrhosis, ↑ risk of hepatocellular carcinoma.

Protein-Losing Enteropathy (PLE)
Loss of protein via gut → edema, effusions, hypoalbuminemia.

Thromboembolism
↑ Risk of clots due to stasis and endothelial dysfunction.

Renal Dysfunction
Reduced perfusion and congestion impair kidney function.

Arrhythmias
Atrial tachyarrhythmias common; can worsen cardiac output.

Clinical Implications in Fontan Physiology

Vasopressor Use
Maintain SVR and optimise preload.
Used to maintain BP and Fontan circulation.

Role of TTE
Assess ventricular function, valves and volume.
Used perioperatively to guide management.

Volume Status
Preload dependent circulation.
Avoid hypo- and hypervolaemia.
Careful fluid management to balance preload.

Sinus Rhythm & PVR
Avoid ↑ PVR (hypoxia, acidosis, pain).
Maintain sinus rhythm.
Close monitoring and optimisation maintained **in all cases.**

Multidisciplinary Care
Essential for planning and perioperative care.
Individualised, physiology-driven approach.

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax*. 1971;26(3):240-248. doi:10.1136/thx.26.3.240
2. Ohuchi H. Adult patients with Fontan circulation: What we know and how to manage adults with Fontan circulation? *Journal of Cardiology*. 2016;68(3):181-189.
3. Nayak S, Booker PD. The Fontan circulation. *Continuing Education in Anaesthesia Critical Care & Pain*. 2008;8(1):26-30. doi:10.1093/bjaceaccp/mkm047

Images are for illustration purposes only. With thanks to all our patients.