The Fontan circulation

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Key points

Increasing numbers of patients with a Fontan circulation are reaching adulthood and requiring anaesthesia for non-cardiac surgery
Understanding the physiology of the Fontan circulation is essential for the successful anaesthetic management of these patients. Anaesthesia should be administered only in a centre where the relevant cardiological and intensive care unit expertise is available.
The older Fontan patient is at particular risk of thromboembolism, arrhythmias and progressive ventricular dysfunction.
Heart function in the pregnant patient with a Fontan circulation deteriorates throughout pregnancy. Elective delivery under epidural anaesthesia is usually the technique of choice.
Laparoscopic and day care surgery is usually well tolerated in the young Fontan patient.

In a normal biventricular heart, the systemic and pulmonary circulations are in series and each circulation is supported by a ventricle. In patients born with a single ventricular chamber, the two circulations are in parallel and patients only survive because the systemic and pulmonary venous bloods mix. In 1971, Francis Fontan and Eugene Baudet first described a procedure that diverted all systemic venous blood into the pulmonary arteries, without the interposition of a ventricle, as a surgical palliation for tricuspid atresia. The introduction of this eponymous ‘Fontan operation’ 36 yr ago revolutionized the treatment of complex congenital heart defects and remains the treatment of choice for patients born with one functional ventricle. A large number of children continue to benefit from the Fontan operation. However, despite many refinements of the surgical procedure in the past 20 yr, a relatively high proportion of patients demonstrate a gradual decline in functional capacity and premature death.

Indications for a Fontan circulation

Conversion to a Fontan circulation is considered in all patients with complex congenital heart disease when a biventricular repair is not possible. These include patients with tricuspid atresia, pulmonary atresia with intact ventricular septum, double inlet left ventricle, hypoplastic left heart syndrome, double outlet right ventricle, and complete atrioventricular septal defects. Selected patients should be in sinus rhythm, have adequately sized pulmonary arteries, and good ventricular function. The absence of any one of these selection criteria increases the risk of a poor outcome.1

Surgical approach

Surgical palliation of a single ventricle is performed in stages. A Fontan circulation is contra-indicated in the neonatal period because of the relatively high (physiological) pulmonary vascular resistance (PVR). Moreover, a staged approach allows progressive adaptation of the heart and lungs and reduces the overall perioperative morbidity and mortality.

Stage 1: systemic-pulmonary shunt

The aim of the initial palliation is to provide complete relief of any systemic obstruction if it exists, and provide pulmonary blood flow just sufficient to allow adequate oxygen delivery to tissues and pulmonary arterial growth. Pulmonary blood flow must be minimized to ensure that PVR is kept low and the ventricle does not have an excessive volume load. This is usually achieved by placing a restrictive synthetic conduit (3–4 mm internal diameter) between a major systemic central vessel and a proximal pulmonary artery (Fig. 1a).

Alternatively, patients with hypoplastic left heart syndrome may have a conduit placed between the right ventricle and the left pulmonary artery, which provides pulmonary blood flow in parallel to systemic blood flow directly from the right ventricle. The major theoretical advantage of this arrangement is the avoidance of aorto-pulmonary runoff, resulting in higher coronary and systemic perfusion pressures and reducing the incidence of ventricular ischaemia.

Stage 2: superior cavopulmonary connection (Glenn type operation)

The second stage palliative procedure consists of a bidirectional Glenn shunt or hemi-Fontan procedure and is usually undertaken as soon as the pulmonary arteries have grown sufficiently to allow a low PVR, usually between 2–6 months. Cardiopulmonary bypass is usually used to allow anastomosis of the superior vena cava to the proximal right pulmonary artery (Fig. 1a). The previous systemic-pulmonary shunt is usually ligated. This intervention provides low-pressure pulmonary blood flow and decreases the volume load on the ventricle. The ventricle continues to receive deoxygenated blood directly from the inferior vena cava (through a large atrial septal defect, enlarged by the surgeon if necessary), in addition to oxygenated blood returning via the pulmonary
veins. This mixing of oxygenated and deoxygenated blood in the functionally single atrium means that the patient will continue to be desaturated, with peripheral oxygen saturations about 80–85%. After surgery, the patient is at risk of developing intrapulmonary arteriovenous shunts, related either to endothelial dysfunction secondary to chronic non-pulsatile pulmonary blood flow or because the lungs are not perfused by some unidentified factor produced by the liver.²

Stage 3: completion of the Fontan circulation

The final stage of Fontan palliation is usually performed at 1–5 yr of age, when restriction of patient activity is becoming problematic and pulmonary arteries are of sufficient size to allow a low PVR. There are a number of different surgical techniques used, but all result in the same basic blood flow pathways. Blood in the inferior vena cava is directed into the pulmonary circuit either via an intra-atrial baffle or, more usually, via an extracardiac conduit (Fig. 1c). Some patients with sub-optimal PVR (10–25%) require a small fenestration (approximately 4 mm diameter) to be created between the conduit and the atrium. This fenestration allows a residual right to left shunt, thereby limiting caval pressure and congestion, and increasing preload of the systemic ventricle and cardiac output, at the expense of slight desaturation.

Most mortality on this staged surgical pathway occurs during and after stage I palliation, with cumulative early and interstage mortality ranging between 5% and 30%. Improved outcome is associated with good patient selection, good preoperative stabilization and perioperative monitoring, and early repair.³

Complications of a Fontan circulation

Although most patients with a Fontan circulation have a good quality of life for many years, others fare less well. The severity and rate of progression of these complications varies between individuals, probably relating to differences in their PVR, ventricular morphology, and atrioventricular valve function.⁴

Diminished exercise tolerance and ventricular dysfunction

All patients with a Fontan circulation have an abnormal cardiopulmonary response to exercise. They have a blunted heart rate response and a limited ability to increase stroke volume with exercise, due to impaired ventricular function and difficulty in increasing ventricular preload. Most 10 yr follow-up studies document myocardial dysfunction and grade II failure in about 70% of patients.⁵ The ventricle becomes dilated, hypertrophic, and hypocontractile, with deterioration in both systolic and diastolic function. In these patients, the dominant limiting determinant of ventricular function is preload, so that inotropes, vasodilators, and β-blockers have relatively little impact.
Arrhythmias

Atrial arrhythmias occur in about 45% of patients in the 10 yr following surgery, the high incidence being related to multiple suture lines near the sinus node, atrial enlargement, and elevated atrial pressure. These are poorly tolerated and can lead to severe haemodynamic deterioration: immediate cardioversion may be necessary. Affected patients require full anticoagulation because of a significant risk of atrial thrombus.

Shunts

Patients with a Fontan circulation, particularly those having a residual right to left shunt through a fenestration, may be slightly desaturated ($S_{O2} \sim 95\%$) even at rest. Drainage of coronary sinus blood into the systemic circulation also contributes to this desaturation. Left to right shunting may occur through aorta-pulmonary collaterals or incomplete occlusion of previous artificial shunts. These shunts result in volume overload on the ventricle and may induce an irreversible increase in PVR secondary to high regional pulmonary blood flow.

Protein losing enteropathy

Protein losing enteropathy is characterized by excessive loss of proteins from serum into the intestinal lumen, possibly due to impedance to drainage of the thoracic duct by high superior caval venous pressure together with mesenteric vascular inflammation. Manifestations include oedema, immunodeficiency, ascites, malabsorption of fat, hypercoagulopathy, hypocalcaemia, and hypomagnesaemia. During a 10-yr follow-up, its incidence is about 13%. The prognosis is poor (60% 5-yr and 20% 10-yr survival after diagnosis). Treatment includes a diet low in salt and high in calories, protein content, and medium chain triglycerides. Diuretics, corticosteroids, heparin, and octreotide (a somatostatin analogue) may provide symptomatic relief in some individuals.

Developmental deficit

These patients are at particular risk of having neurological and developmental deficits due to the impact of multiple episodes of cardiopulmonary bypass, thrombotic events, chronic hypoxaemia, and pre-existing neurological deficits.

Thromboembolism

About 30% of patients have thromboembolic events, according to most 10-yr follow-up studies. A low flow state, atrial scarring, arrhythmias, dehydration, and a hypercoagulable state all increase the risk of thromboembolism. Prevention of pulmonary embolism is essential; hence, prophylactic anticoagulation with warfarin or anti-platelet agents is always prescribed.

Anaesthetic management

A full understanding of the physiology of the Fontan circulation is essential for ensuring optimal anaesthetic management of these patients. The main determinants of the success of the Fontan circulation are systemic venous pressure, PVR, atrioventricular valve function, cardiac rhythm, and ventricular function. Disturbance of any of these factors compromises cardiac output.

The driving force for blood flow through the pulmonary circulation is the difference between central venous pressure (CVP) and atrial pressure. There is no active pumping of blood through the lungs. Intravascular volume is the main determinant of CVP; therefore, hypovolaemia is tolerated poorly. Sinus rhythm is one of the three important factors, along with atrioventricular valve and ventricular function, in maintaining optimal ventricular filling and atrial emptying.

Drugs that have negative inotropic effects (e.g. β-blockers) should be prescribed with caution. Similarly, drugs that increase PVR (e.g. α-adrenergic agonists) should be avoided. As with all patients, factors that increase PVR (e.g. alveolar hypoxia, hypercarbia, acidosis) should be strenuously avoided.

Preoperative assessment

The functional status and comorbidities found in these patients varies significantly, from the young patient who is well compensated to the adult with a failing ventricle. A detailed history, physical examination, and baseline haematological and biochemical investigations are always necessary, even before minor surgery. End-organ damage may be present, secondary to low cardiac output and chronically high venous pressure. A 12-lead ECG and echocardiography allow assessment of rhythm, and ventricular and valvular function. Perioperative antibiotic prophylaxis with broad spectrum cover is required for all procedures likely to produce a bacteraemia.

The risk of air or fat emboli occurring during major surgery is relatively high in patients with a fenestration. Some cardiologists recommend closing a fenestration preoperatively, using devices introduced percutaneously, for patients undergoing high-risk surgery. Appropriate equipment and trained personnel should be available to handle the potential challenges posed by these patients.

Monitoring

In addition to monitoring of oxygenation, gas analysis, and cardiac rhythm, invasive arterial and CVP monitoring is mandatory in Fontan patients undergoing major surgery, particularly where significant volume shifts are likely to occur. Monitoring of trend CVP can help in the assessment of vascular volume status, though it reflects only mean pulmonary artery pressure (mPAP), not ventricular preload. Typical pressures in a well compensated young patient with a Fontan circulation are a CVP (mPAP) of 12 mm Hg.
Intrathoracic pressure. This causes decreased venous return, decreased pulmonary blood flow, and hence, decreased cardiac output. Low respiratory rates, short inspiratory times, low PEEP, and tidal volumes of 5–6 ml kg\(^{-1}\) usually allow adequate pulmonary blood flow, normocarbia, and a low PVR. Hyperventilation tends to impair pulmonary blood flow, despite the induced respiratory alkalosis, because of the increased mean intrathoracic pressure.

**Induction and maintenance**

Induction of anaesthesia can reduce cardiac output, secondary to myocardial depression, systemic vasodilatation, and artificial ventilation. It is advisable to avoid induction agents that depress myocardial contractility (e.g. thiopental). The transient systemic vasodilatation caused by propofol is usually less problematic, as long as normovolaemia is ensured.

High concentrations (>1.5 MAC) of volatile anaesthetic agents should not be used to maintain anaesthesia, as they increase the likelihood of arrhythmias. Instead, a low concentration of an inhalational agent in combination with a short-acting opioid (e.g. remifentanil) provides a cardiosable anaesthetic. Inspired oxygen concentration should be adjusted so that the peripheral oxygen saturation is >95%. An increase in oxygen requirements is indicative of increasing right to left shunting through a fenestration or intrapulmonary shunts. This will be due to a decrease in ventricular function, decreased pulmonary blood flow, ventilation-perfusion inequalities, or any combination thereof. Inadequate pulmonary blood flow may occur either secondary to decreased vascular volume or increased PVR. The most common causes of a peroperative increase in PVR are inadequate analgesia or anaesthesia, hypercarbia, acidosis, use of vasoactive drugs, and increased mean intrathoracic pressure.

Fluid administration must be guided by CVP, TOE, or oesophageal Doppler. Even moderate hypotension may be damaging in Fontan patients, because it may result in a critically low arteriovenous pressure gradient, resulting in inadequate tissue perfusion, a metabolic acidosis, and increase in PVR. Regional anaesthesia, although it does not directly affect PVR or ventricular function, has the potential to produce relative hypovolaemia. Adequate monitoring and appropriate fluid administration should prevent this.

**Mechanical ventilation**

For relatively short procedures, Fontan patients are probably better off breathing spontaneously, as long as severe hypercarbia is avoided. For major surgery, or when prolonged anaesthesia is required, control of ventilation and active prevention of atelectasis is usually advisable. Potential disadvantages of mechanical ventilation in Fontan patients relate to the inevitable increase in mean intrathoracic pressure. This causes decreased venous return, decreased pulmonary blood flow, and hence, decreased cardiac output. Low respiratory rates, short inspiratory times, low PEEP, and tidal volumes of 5–6 ml kg\(^{-1}\) usually allow adequate pulmonary blood flow, normocarbia, and a low PVR. Hyperventilation tends to impair pulmonary blood flow, despite the induced respiratory alkalosis, because of the increased mean intrathoracic pressure.

**Postoperative care**

Adequate monitoring of vascular volume status after major surgery, particularly when further fluid shifts are likely to occur, requires the patient to be nursed in an intensive care unit. Patient controlled intravenous or epidural opioid analgesia help provide good quality pain relief after major surgery and minimize sympathetic tone. Monitoring of oxygen saturations should be performed in all patients for at least 24 h after major surgery, and the inspired oxygen concentration adjusted to maintain saturations above preoperative levels. Thromboprophylaxis should be continued throughout the perioperative period. Patients taking warfarin and undergoing minor surgical procedures may need hospital admission for a short time so that the period of reduced anticoagulation is kept as short as possible.

Young Fontan patients in good health may have minor surgery performed as a day case procedure, if they have no specific contra-indications. Attendance at a pre-admission clinic the week before surgery is advisable. Routine postoperative care should be supplemented by continuous oxygen saturation monitoring until such time as they fulfil certain discharge criteria (Table 1).

**Pregnancy and delivery**

The number of patients with a Fontan circulation reaching childbearing age increases every year, though infertility and miscarriage are common. Although women can successfully complete pregnancy with a Fontan circulation, the associated physiological haemodynamic changes have a significant influence on their abnormal heart function. Even by 20–28 weeks gestation, myocardial oxygen consumption and heart rate normally increase by 20% and stroke volume by 40%, the latter related to a physiological increase in blood volume. This fluid retention may result in atrial distension, which may in turn trigger atrial tachyarrhythmias and significant haemodynamic disturbance.

<table>
<thead>
<tr>
<th>Table 1 Minimum discharge criteria for Fontan patients after day care surgery</th>
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<td>Oxygen saturations in air maintained at preoperative values</td>
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<tr>
<td>No bleeding</td>
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<tr>
<td>Excellent pain control</td>
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<td>No nausea or vomiting</td>
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<td>Patient drinking normally</td>
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<td>Adequate home support from accompanying carers</td>
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<td>Carers have own transport</td>
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<td>Patient lives within 30 min travelling time of hospital</td>
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Provision of anaesthesia for pregnant patients with a Fontan circulation follows the general principles outlined above, i.e. avoidance of factors that increase PVR, maintenance of ventricular function, and avoidance of hypovolaemia. Most pregnant women with a Fontan circulation will be fully anticoagulated, as both pregnancy and their abnormal cardiovascular anatomy make them more likely to suffer thromboembolic complications. Women on warfarin should be converted to heparin in late pregnancy. An elective delivery is preferable as it enables anticoagulant therapy to be stopped for the minimum time before an epidural catheter is inserted. Epidural analgesia, perhaps combined with an intrathecal opioid, is the technique of choice for the provision of analgesia both during elective labour and surgery. Excessive straining during the second stage of labour, which reduces pulmonary blood flow via its effects on intrathoracic pressure, should be avoided and a forceps assisted delivery is usually advisable. High neuraxial blocks may lead to significant vasodilatation and relative hypovolaemia, which may decrease pulmonary blood flow and ventricular preload. Gradual titration of epidural analgesia, appropriate fluid loading, and left uterine displacement help avoid sudden hypotension and the need for vasoconstrictors. Vasopressors with α-agonist activity should be avoided as they increase PVR. CVP monitoring is essential in the event of haemodynamic instability, such as a heavily bleeding patient.

**Laparoscopic surgery**

The potential advantages of laparoscopic surgery include reduction of postoperative pain and recovery time. The potential complications of the induced pneumoperitoneum include hypercarbia, gas embolism, haemodynamic disturbance, pneumothorax, and mediastinal emphysema. Using an intra-abdominal pressure of <10 mm Hg, there is increase in venous return with subsequent increase in cardiac output. In contrast, if the intra-abdominal pressure exceeds about 15 mm Hg, compression of the inferior vena cava results in a decrease in venous return and cardiac output. Carbon dioxide absorption and impaired ventilation due to abdominal distension may result in hypercarbia and, subsequently, increased PVR. The presence of a fenestration in the Fontan circulation increases the risk of paradoxical carbon dioxide embolism. Despite these potential problems, most young patients with a Fontan circulation tolerate laparoscopic abdominal surgery very well, as long as the intra-abdominal pressure is kept at ≤10 mm Hg, the duration of surgery is minimized, and adequate ventilation and intravascular volume are maintained.9

### References

4. Jacobs ML, Pelletier G. Late complications associated with the Fontan circulation. *Cardiol Young* 2006; 16(Suppl 1): 80–4
5. Walker F. Pregnancy and the various forms of the Fontan operation. *Heart* 2007; 93: 152–4

Please see multiple choice questions 21–25