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A review of mechanical ventilation strategies in children following the Fontan procedure

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Abstract

This article briefly outlines the development of the Fontan operation, and details ventilation strategies after such surgery.

MeSH: Fontan procedure, Thoracic surgery, Ventilation

The introduction of the Fontan procedure 30 years ago revolutionized the treatment of complex congenital heart defects where biventricular repair is not possible. Fundamental to the physiology of the Fontan circulation is dissociated from a ventricular power source and the pulmonary vascular resistance is in series with systemic vascular resistance.

Much has been written about the procedure itself, but most studies and series of Fontan patients have dealt with patient selection and operative technique. Few studies have focused specifically on perioperative and post-operative management of these patients. Additionally, any mention of mechanical ventilation that may be required in the post-operative management usually revolves around early extubation when possible. The purpose of the current review is to discuss aspects of the post-operative management specifically relating to the ventilatory management of these patients.

Surgical Perspective

In 1971, Francis Fontan and Eugene Baudet first described a procedure that diverted all systemic venous blood into the pulmonary arteries as a surgical repair for tricuspid atresia[1]. In their initial description, Fontan recognized some of the post-operative challenges that continue in the present day. He described the need to "provide a large amount of fluid to ensure a correct hemodynamic balance", advantages to low intrapulmonary pressures, early extubation, as well as the tendency to develop pleural effusions. These factors continue to impact the post-operative morbidity and mortality of patients despite advances on Fontan's original operative procedure.

Fontan's first description of the procedure consisted of connecting the superior vena cava to the distal right pulmonary artery, and the right atrium to the proximal right pulmonary artery; thereby the right atrium was "ventriclized". Many authors have since proposed alterations to the procedure, but there have been two most notable.

Hopkins and associates introduced the bi-directional Glenn shunt in 1985. This aided DeLeval in the development of total cavopulmonary anastomosis utilizing his lateral tunnel technique with the belief that there would be less turbulence and improved venous return in bypassing the right atrium entirely. Additionally, this modification has the advantage of eliminating atrial suturing lines, thereby decreasing the incidence of post-operative atrial arrhythmias." It has also been suggested that this allows the right atrium to avoid being subjected to elevated central venous pressures.

Finally, a recent modification involves the fenestration of the atrial septum. Its use has been advocated in the setting of high-risk patients who may not have the ability to tolerate diversion of the entire systemic venous return to the pulmonary circuit. The fenestration may be either adjustable during the post-operative period or closed later in the catheterization laboratory. The main physiologic advantage proposed by groups employing these techniques is the ability to increase cardiac output with minimal reductions in systemic saturation. After a transitional period, patients may no longer require or benefit from the persistence of a vent. Some disadvantages to fenestration are that some patients have significant hypoxemia and have polycythemia gradually develop. Other patients maintain an adequate degree of oxygenation but may have heightened risk of embolic complications.

Preoperative Risk Assessment

Current risk factors for the Fontan procedure have been outlined in several large series.' (Table) Since pulmonary blood flow will be determined by the resistance across the pulmonary bed post-operatively, the pulmonary vascular resistance and the anatomy affecting it are important considerations. Elevated pulmonary vascular resistance to greater than 4 Wood's units is a specific risk factor for death in the perioperative phase.

Table Preoperative risk assessment in candidates for the Fontan operation. PVR, pulmonary vascular resistance

Variable	Low risk	Medium risk	High risk
PVR (Wood's units)	<2	2-4	>4
Mean PAP (mmHg)	<15	15-20	>20
LVEDP (mmHg)	<8	8-12	>12
EF (%)	>60	45-60	<45

PAP, pulmonary artery pressure; LVEDP, left ventricular end-diastolic pressure; EF, ejection fraction.

An anatomic factor to consider is the size of the pulmonary arteries. Small pulmonary arteries increase the risk of complications with the Fontan procedure. The exact size associated with an increased risk is not known. Attempts have been made to classify the risk with a ratio of pulmonary artery size to aorta size. The Fontan procedure may still be performed in those with small pulmonary arteries if they are augmented during the procedure. Additionally, arborization anomalies and the size of the vascular bed and its intrinsic resistance are fundamental factors often disturbed if the central pulmonary arteries are small.

Ventricular function, particularly diastolic function, is also important to a successful Fontan procedure. Ventricular hypertrophy secondary to volume overload or outflow track obstruction decreases left ventricular compliance. Poor left ventricular compliance increases both left ventricular end diastolic pressure and left atrial pressure. These changes affect the perfusion pressure gradient for inflow into the left ventricle. This can result in an increased mean pulmonary pressure and central venous pressure after the Fontan operation.

Postoperative Care

Circulation

Post-operatively, patients have surgically altered pathways for systemic venous return that significantly affects their cardiovascular physiology. Pulmonary blood flow and therefore cardiac output, is dependent on passive, non-pulsitile flow of returning systemic venous blood. The goal of post-operative management is to optimize cardiac output at the lowest central venous pressure possible. In order to understand the impact of these factors on the post-operative course of a Fontan patient, some physiologic principles must first be discussed.

As previously stated, pulmonary blood flow is no longer determined by the pumping of the right ventricle. Instead, there is reliance upon an energy gradient between the systemic veins and the pulmonary artery without a pressure gradient. Pre-operative factors affecting this gradient will be problematic during the post-operative course and must be clearly communicated to those participating in the patient's management.[14] Those with elevated pulmonary artery pressures may not be acceptable candidates for the procedure as the systemic venous pressure may be unable to overcome the resistance and provide forward blood flow[17]. Thus, the Fontan is not performed in the neonatal period due to the presence of normally high pulmonary vascular resistance.

The fluid status of these patients must be closely monitored to ensure an adequate central venous pressure as the driving force. Venous return and pulmonary blood flow are maintained through adequate intravascular volume repletion and patient positioning. Blood return from the superior vena cava is aided by gravity. Simple interventions such as having the patient sit up or elevating the head of the bed will improve return. Similarly, elevating the legs may augment blood return from the inferior vena cava.

In 1981, Heck and Doty proposed phasic external lower body compression as an adjunct to assist circulation in the Fontan patient. It was their feeling that this technique diminished fluid sequestration and improved cardiac output. The use of this technique by the use of mast trousers has, in recent years, been abandoned for the approach of early extubation in the post-operative period.

Ventilator Management

Elevated intrathoracic pressures can affect the pressure gradient for venous return. Such elevations in intrathoracic pressures commonly occur with positive pressure mechanical ventilation. Spontaneous respiration creates a negative intrathoracic pressure with inspiration, which increases systemic venous return. If spontaneous respiration can be achieved without pain and subsequent splinting, it is understandable that hemodynamics would be improved.[3]

End-Expiratory Pressure

The application of end-expiratory pressure to the Fontan patient can have a significant negative effect on the circulatory state. However, the application of an “appropriate” level of end-expiratory pressure has substantial beneficial effects, especially in Fontan physiology. The level of “appropriate” positive end-expiratory pressure (PEEP) for lung disease in the Fontan patient remains unknown. Howell et al demonstrated that in the non-Fontan patient pulmonary vascular resistance may fall at low levels of PEEP with subsequent increases at higher levels of PEEP. In contrast, Williams et al demonstrated in Fontan patients that pulmonary vascular resistance is increased at all levels of PEEP (3 to 12 cm H_2O), and the cardiac index is decreased at high levels of PEEP (9-12 cm H_2O).

PEEP maintains functional residual capacity and increases PaO_2 after the Fontan procedure, which is a beneficial affect for the patient. Maintenance of functional residual capacity leads to a decrease in pulmonary vascular resistance by avoidance of atelectasis and hypoxic vasoconstriction. Specifically related to cardiac surgery, PEEP has been demonstrated to have beneficial effects on arterial oxygenation, atelectasis, and right-to-left shunting in children after a variety of cardiac operations.

Evolution to Early Extubation

There has been a progressive move toward the early extubation of Fontan patients in the post-operative period.[3] Spontaneous respiration creates a negative intrathoracic pressure with inspiration, increasing systemic venous return. The concern is splinting secondary to pain will result in atelectasis, and thus worsen hemodynamic performance. This must be balanced with the effects of increased intrathoracic pressures caused by positive pressure ventilation. These pressures have been shown to decrease diastolic flow into the pulmonary arteries in these patients. Studies have also shown improved pulmonary blood flow and cardiac output with spontaneous respiration after the Fontan and are now promoting early extubation, either in the operating room or on arrival to the ICU.[3]

Alternative Ventilation Strategies

High Frequency Ventilation

There have been individual reports and small series examining the use of high frequency ventilation in Fontan patients with significant lung injury. Overall, these and others have concluded that while caution is necessary in Fontan physiology, patients have tolerated high frequency ventilation with minimal discernable adverse hemodynamic effects.[35]

Negative Pressure Ventilation

Negative pressure ventilation has been proposed for use when the post-operative course is complicated by a low cardiac output state.[31] Shekerdemian et al demonstrated an increase in cardiac output and improved hemodynamic status with negative pressure ventilation as compared to positive pressure ventilation. However, to date, negative pressure ventilation has not gained widespread use.

Non-invasive Ventilation

The use of noninvasive forms of ventilation in the postoperative Fontan patient has some theoretically intriguing possibilities. By combining spontaneous respiration with some positive distending pressure, there exists the possibility of combining “the best of both worlds.”

Noninvasive ventilation, in the form of continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BiPAP), have been previously examined in the setting of post-operative cardiac surgery patients. Additionally, both forms of ventilation have been used in the therapy of congestive heart failure patients. In these patients, noninvasive ventilation appears to have an impact on cardiac output and venous return, though possibly to a somewhat lesser degree than in intubated patients. However, there has yet to be a study examining the differences in Fontan physiology.

Inhaled Nitric Oxide

The use of a selective pulmonary vasodilator in the Fontan patient is desirable. The introduction of inhaled nitric oxide (NO) has allowed the avoidance of the systemic vascular resistance decline found with non-selective, systemic pharmacotherapy such as priscaline. The use of NO has been well studied in the setting of pulmonary hypertension in the neonate suffering from meconium aspiration syndrome. However, the use of NO in the post-operative cardiac surgery patient remains unstudied. To date, there is a lack of controlled studies demonstrating that its use has an impact on important variables such as length of mechanical ventilation, length of hospital stay, or mortality. Additionally, the balance between the desire for early extubation and the theoretical advantages to NO delivery must be weighed. Widespread trials of NO delivery via nasal cannula or facemask are only now being undertaken.

Summary

Post-operative management of the Fontan patient remains a demanding exercise. This fact is compounded by the lack of randomized, controlled trials examining various ventilator strategies. Clinicians are forced to extrapolate from animal models, case series, or individual experiences. What is known is that striking the proper balance between the beneficial and detrimental effects of mechanical ventilation in the Fontan patient remains challenging. The ventilatory management of these patients requires an individualized approach.

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