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Patient With a Failing Fontan for Emergency Exploratory Laparotomy

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Stem Case and Key Questions Content

The patient, 24 years old with PMH of Hypoplastic Left Heart Syndrome (HLHS) (Fig. 2-A from selected reference # 1), had a Norwood operation and a Blalock-Taussig shunt (Fig. 2-B, from selected reference # 1) at 2 weeks of age followed by a classical Glenn shunt at the age of 8 months and a Fontan procedure at the age of 4 years. Three years ago he was presented with increased fatigability, atrial arrhythmias and an oxygen saturation of 75 %. He underwent a Fontan revision and coil embolization of pulmonary arterial venous malformations (AVM). Following Fontan revision surgery he noted subjective improvement and his oxygen saturation improved to 96 %. However, he suffered from renal insufficiency post-operatively. He currently is fatigued with minimal exertion and experiences transient episodes of tachyarrhythmias.

QUESTIONS:
1. Describe briefly Fontan anatomy and physiology. Name congenital heart defects which required Fontan as the last step of palliative correction.
2. What is the long-term outcome of a Fontan procedure?
3. What is classical Glenn and bi-directional Glenn shunt? What do you think was the original Fontan modification and what type of revision has the patient had? What are the differences between a *Lateral Tunnel* and an *Extracardiac* Fontan? What is the purpose of a *fenestration*?
4. How do you explain low oxygen saturation before the last surgery and post-operative improvement?

CASE PRESENTATION

The patient is going to have exploratory laparotomy for a small bowel obstruction. Physical exam is significant for mild ascites; BP 100/50, HR 100, RR 22; lab: Hct 29, Plt 158, Abumin 2.0 and slightly elevated liver enzymes, PT, PTT normal.

QUESTIONS:
5. What is the presentation of Fontan failure?
6. What factors contributed to the failure?
7. What conditions will eventually require heart transplant and what can be corrected by other means?
8. What diagnostic modalities are utilized in the assessment of Fontan failure patients?
9. What is the effect of Fontan physiology on the liver and splanchnic circulation?
10. What is your plan for monitoring? Do you plan any invasive monitoring? What are the risks

and benefits of invasive monitoring?
11. What is your plan for anesthesia induction and maintenance?

CASE PRESENTATION
You proceed with the case after placement of a preinduction arterial line. Induction and intubation are uneventful; however, while the case is progressing you start to experience difficulty in maintaining the patient's systemic blood pressure.

QUESTIONS:
10. What is your differential diagnosis of hypotension?
11. How will you adjust your management?
12. What CVP constitutes adequate preload for Fontan patients?

CASE PRESENTATION
The procedure is almost done. You received a recently sent blood gas, which showed: 7.26/32/70/-6/18/93%, lactic acid 3.3. The surgeon is planning to place the patient in ICU and asks you if he should remain intubated.

QUESTIONS:
13. What are the warning signs you see on the ABG?
14. What is the effect of respiration on splanchnic venous flow and pulmonary arterial blood flow?

Model Discussion Content

Fontan physiology.

- The Fontan procedure is the last step in palliative correction for congenital heart lesions with a single ventricle (right, left, or indeterminate morphology) (Fig. 2-A, 1-A from selected reference 1).The purpose of this correction is to dedicate the single ventricle to the systemic circulation and to provide pulmonary circulation by diverting systemic venous return to the pulmonary artery, excluding the RV as a pump to the pulmonary circulation (selected reference 2, 3). Unfortunately, the role of the RV in maintaining a low central venous pressure and preventing the sequestration of blood volume in the very distensible systemic venous system is irreplaceable (1).

- Pulmonary blood flow occurs passively as a result of a systemic venous to pulmonary venous pressure gradient. Pulmonary perfusion depends on an adequate systemic venous return (CVP pressure), low PVR and normal systolic and diastolic function of the single ventricle.

- Cardiac output is preload-limited, and limited by pressure or flow resistance across the pulmonary vascular bed. Fontan circulation leads to contractility-afterload mismatch by means of increased impedance caused by additional connection of the pulmonary circuit. This adds a 10 % increase to total peripheral vascular resistance. Thus, the systemic and pulmonary vascular resistance has to be overcome in series. This situation reduces ventricular mechanical

efficiency and leads to a limited cardiac functional reserve.

Long-term outcome of the Fontan procedure.
The incidence of early failure has dramatically decreased over the past several years and is related to modification of both the selection criteria and the operative techniques. However, a decline in survival continues after 5 - 7 years (2 - 4).

Fontan and Glenn modifications.
Historically during a classical Glenn, the superior vena cava (SVC) was disconnected from the right atria and connected to the right pulmonary artery, while the continuity of the right and left pulmonary artery was surgically interrupted. Glenn shunts do not completely correct cyanosis because flow from the inferior vena cava (IVC) still enters the systemic circulation. As patients grow, flow from the IVC increases reducing systemic saturation. Over time venovenous collaterals from the SVC to the IVC reduce flow through the Glenn shunt. During the following Fontan procedure, the right atrium was connected to the left pulmonary artery. The reason for cyanosis after such modification of a Fontan procedure might be the arterial venous malformations (AVM), which are believed to develop secondary to the lack of the hepatic hormonal factor delivered from the venous circulation to the pulmonary artery and subsequently equally to both lungs. A patient who had a classical Glenn with the pulmonary artery divided and the SVC connected to the right pulmonary artery, followed by the Fontan procedure connecting the right atrium to the left pulmonary artery, will be prone to development of such AVM. A Fontan revision with the reestablishment of right and left pulmonary artery continuity, diverting all the venous flow including hepatic return to both lungs should improve saturation. Three years ago this patient had such a revision with conversion to an extra-cardiac Fontan and coil embolization of the AVM. In modern modification of the bi-directional Glenn (BDG) procedure, the SVC is connected to the ipsi-lateral PA permitting blood to flow into both the right and left pulmonary arteries (Figure 1-C from selected reference 1). Having the BDG procedure before completion of Fontan allows a decrease to the diastolic dysfunction, which is common in the SV patient (5). The following Fontan procedure includes the anastomosis from the IVC to the PA. The IVC-PA connection is completed either as an intra-atrial lateral tunnel through the atrium (Fig 3-B from selected reference 1) or as an extracardiac conduit to the pulmonary artery (Fig. 3-C from selected reference 1). Sometimes 4 mm fenestration will be placed between the conduit and atrium. The fenestration will lead to some venous admixture with oxygenated blood, but it decreases the incidence of early Fontan failure by permitting adequate volume loading of the ventricle. Fenestration has not been shown to impact late outcomes after Fontan palliation (4).

Presentation of Fontan failure.
- Progressive exercise intolerance.
- Incapacitating atrial tachyarrhythmias.
- Unremitting pleural effusion.
- Cyanosis secondary to AV malformations.
- Protein losing enteropathy (PLE).

Factors contributing to Fontan failure.Fontan failure is often multi-factorial.

Correctable causes:
- Atrial arrhythmia: Atrial fibrillation in patients with a single ventricle physiology will be much less tolerated than in a two-ventricle population.
- AV valve regurgitation.
- Pulmonary venous compression.
- Aortopulmonary collaterals, pulmonary arteriovenous malformations.
Uncorrectable causes, which require a heart transplant (6):
- Ventricular dysfunction. After systemic to pulmonary shunt (BT-shunt) (Figure 1-B and 2-B from selected reference # 1), a single ventricle was working for pulmonary and systemic circulations in parallel. This situation predisposes the ventricle to volume overload, ventricular hypertrophy and diastolic dysfunction. After Fontan completion, the single ventricle supplies systemic and pulmonary circulations in series. The ventricle is chronically volume underloaded and this promotes diastolic dysfunction.

A Glenn procedure preceding the Fontan procedure reduces ventricular volume overload, gives time for ventricular hypertrophy to subside, and improves ventricular contractility. After load increases after both stages, which can cause fatal after load mismatch in high-risk patients. This after load increase is smaller, if the procedure is done in stages, and the after load decrease occurs during the interval between the procedures. Thus, a staged procedure results in an improvement in ventricular efficiency (5).

The cardiac index of Fontan patients' decreases compared to bi-ventricular patients in spite of similar systemic blood pressure, ventricular contractility, and diastolic stiffness. Lack of the right ventricle results in higher ventricular work (higher power expenditure) per unit of forward flow.
- Pulmonary vascular diseases have multifactorial etiologies. Failure to protect the pulmonary vascular bed early in life is one of them. Another one is the absence of the RV pumping force; the lower energy pulmonary circulation increases the pulmonary vascular impedance and the pulmonary afterload. The pulmonary vascular resistance index is higher in Fontan patients compared with bi-ventricular patients.

Others causes are:
- Increased pulmonary lymphatic pressure.
- Thromboembolic event.
- Elevated left atrial and pulmonary venous pressure.
Corrections (7):
- Reoperation:convert atriopulmonary and atrioventricular connections to extra-cardiac Fontan (more energy-efficient)
- Fenestration
- Ablation of atrial arrhythmias
- Correction of atrioventricular valve regurgitation
- Correction of subaortic obstruction

- Heart transplantation
Primary indication for transplantation - ventricular dysfunction (6):
Diagnostic modalities:
- Echocardiography.
Quantification of ventricular function; EF is subjective in the setting of right ventricular morphology.
- Magnetic resonance imaging (MRI) provides greater accuracy of EF than echocardiography.
- Cardiac catheterization.

Ventricular end-diastolic pressure (VEDP) - objective index of ventricular function. More than 12 mmHg indicates significant dysfunction.
Quantification of PVR (not reliable).
Elimination of large aorto-pulmonary collaterals can also be accomplished also during catheterization.
Preoperative evaluation and anesthetic management.
During the preoperative evaluation, it is very important first to realize what is the congenital pathology and to what degree palliation has been completed at the time of assessment. Because SV ventricle palliative procedures have developed and have been significantly modified over time, the exact nature of the connections created has to be understood and the cardiologist should assess their patency.
Information regarding the integrity of
- Valve function,
- PVR,
- Ventricular function,
- Ventricular end-diastolic pressure,
- Collateral circulation,
- Presence of uncoiled AVMs
Laboratory tests should include
- Complete blood count,
- electrolytes,
- BUN, Cr,
- coagulation profile,
- Liver function tests.

Anesthetic management is directed to preserve ventricular function and promote pulmonary blood flow. Hypovolemia is poorly tolerated because it significantly reduces pulmonary blood flow, ventricular preload, and output. Volume status of the patient with an acute abdominal problem is a special concern. Anesthetic induction often produces vasodilatation requiring volume administration. Impaired ventricular function may respond to inodilator therapy with milrinone. Invasive hemodinamic monitoring as arterial line and CVP line is helpful in perioperative management; however, access may be difficult due to prior multiple surgeries and catherization. Also the risk of arterial and venous thrombosis with indwelling catheter (especially in Fontan pathways) has to be weighted against the benefit of monitoring. CVP line inserted in

the internal jugular vein in Fontan patients reflects the mean PA pressure, while in the femoral vein it reflects more central venous pressure. The patient's position and abdominal pressure will affect the measurements. High left atrial pressure secondary to ventricular dysfunction or valvular abnormality will also affect CVP pressure. Preferably CVP is about 14 mmHg, but in many patients a higher CVP pressure (18 - 20 mmHg) may be required to maintain blood pressure and cardiac output.

Transesophageal echocardiography can help in monitoring of ventricular function and volume status, but requires expertise of pediatric cardiologist.

Ventilatory management is directed to promote maximal pulmonary blood flow by minimizing mean airway pressure and providing increased expiratory time. In Fontan patients on mechanical ventilation, hypotension can be caused by decreased preload secondary to increased PVR and inadequate pulmonary blood flow. Causes of increased PVR: light anesthesia, hypoxia, hypercarbia, acidosis, vasoactive drugs, increased airway pressure. Adjust management to decrease PVR: increase anesthesia depth, change ventilator setting to decrease airway pressure (to keep mean airway pressure below 15 cm H2O): large TV, slow RR, I: E ratio 1:3 or 1:4 (pulmonary flow happens mostly during exhalation, so increased expiratory time is recommended). Spontaneous ventilation with negative airway pressure will decrease PVR, increase pulmonary blood flow and increase cardiac output.

Postoperative Management
Most Fontan patients will benefit from recovery in the intensive care unit, unless very minimal surgery was done. The patient in this case presentation with extensive abdominal surgery and metabolic acidosis is definitely an ICU candidate. However, the decision about postoperative mechanical ventilation is not so simple. It will help to maximize oxygen delivery by providing higher FiO2, optimal tidal volume, decreasing work of breathing. In case of respiratory distress syndrome it can improve oxygenation by providing PEEP. Disadvantages of mechanical ventilation: airway infections, lung trauma, decrease in preload with higher PEEP, requirement for heavy sedation. Positive airway pressure increases PVR, decreases pulmonary blood flow, increases shunt and decreases cardiac output. Spontaneous respiration with negative airway pressure will decrease PVR, increase pulmonary blood flow and increase cardiac output. Whether to extubate patient or keep him on mechanical ventilation is a risk/benefit decision. Intravenous or intramuscular narcotics are used routinely after surgical procedure; however, risk of respiratory depression has to be kept in mind in preparation for extubation. Non-steroidal anti-inflammatory agents (ketorolac IV or IM) and local anesthetic infiltration have to be considered in combination.

Effect of respiration on venous and pulmonary blood flow.
Patients with an atrio-pulmonary (AP) connection have phase venous flow affected by the cardiac cycle (atrial reversal, systolic and diastolic flow). Patients with total cavopulmonary connection (TCPC) have a more continuous flow pattern independent of the cardiac cycle, but more dependent on the respiratory cycle (increased flow during inspiration and decreased

during expiration). The absence of atrial reversal in TCPC patients protects hepatic circulation and is probably responsible for decreased incidence of ascites and PLE. Portal venous flow on the other hand increases during expiration. However, in Fontan patients with poor functional conditions, portal venous flow loses the normal expiratory augmentation. These suboptimal flow dynamics, coupled with higher splanchnic venous pressures and a lower transhepatic venous pressure gradient, may be responsible for late gastrointestinal problems.

Effect of Fontan physiology on liver and splanchnic circulation (8).
Elevated central venous pressure in Fontan patients is a recognized as being an inevitable consequence. The SVC venous system is not as dispensible as the IVC system, particularly the splanchnic circulation. Elevated venous pressure is transmitted to the hepatic and portal venous systems and causes a decreased transhepatic gradient, hepatic venous congestion and ascites. Hepatic sinusoidals are highly permeable to proteins; an elevation in hepatic vascular pressure will lead to excessively high lymphatic filtration through the thoracic duct. Since the impedance to systemic lymphatic outflow is increased in Fontan circulation, lymphatic flow will find a low-resistance path to the peritoneal cavity forming ascites or will spill protein-rich lymph into the gut lumen (PLE).

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