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Case Report

Anesthetic management of a single-ventricle pediatric patient with scoliosis: A case report

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ABSTRACT

Single ventricle is a complex cardiac disease which carry high morbidity and mortality and the average 3 years survival rate without Fontan procedure was approximated to be 75%. The most common etiologies are: hypoplastic left heart syndrome and tricuspid atresia. When those patient plan to go for surgery, it's very essential for anesthetists to understand how the physiology is affected rather than etiology. BT shunt patency, right ventricular function and pulmonary artery pressure should be evaluated before any procedure under anesthesia. Scoliosis corrective surgery is a procedure that has its special considerations related to restrictive pulmonary disease, prone positioning, and major bleeding that may occur, which altogether will add to the risk when being conducted in a single-ventricle patient and will necessitate vigilance and special care during monitoring intraoperatively. Here, we report a simple way to monitor the transpulmonary pressure gradient through two central lines in the femoral and jugular veins as part of the patient's anesthesia management.

1. Introduction

Single ventricle heart defect represents 7% of all congenital heart diseases with an approximate incidence 7 of every live birth.[1–3] The two most common forms are: tricuspid atresia and left hypoplastic heart syndrome.[4] It is essential that anesthetists understand how the physiology is affected in that defect rather than the etiology. BT shunt patency, RV function, and pulmonary artery pressure should be evaluated before any procedure under anesthesia; elective and semi elective procedures should be postponed until reaching a more resilient physiology as the ventricle in this stage is overloaded.[5] At the age of 3–6 months, the Glenn procedure is usually conducted.[6] It is the second step of the staged repair for single-ventricle patients; systemic venous blood is directed from superior vena cava to pulmonary circulation, making the ventricle partially unloaded and the physiology more resilient and therefore more tolerant to most the major procedures under anesthesia with special consideration.[6] Scoliosis corrective surgery is a procedure that has its special considerations related to restrictive pulmonary disease, prone positioning, and major bleeding that may occur, which altogether will add to the risk when being conducted in a single-ventricle patient and will necessitate vigilance and special care during monitoring intraoperatively.[7] Here, we report a simple, yet, unique way to monitor the transpulmonary pressure gradient through two central lines in the femoral and jugular veins during

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the management of a single-ventricle patient who underwent scoliosis corrective surgery.

2. Case History

We report helpful monitoring of the pressure gradient between the common atrial pressure and the pulmonary pressure through two central lines in the femoral and jugular veins during the management of a single-ventricle patient who underwent scoliosis corrective surgery and hemivertebra excision of the thoracic spine with its unique problems. Our patient is a 4-years-old male with a history of tricuspid atresia and transposition of the great vessels (TGA) with the unrestrictive pulmonary flow, hypoplastic left aortic arch, apical ventricular septal defect (VSD), and large PDA, who had Glenn shunt. He had scoliosis along with kyphosis; thoracic MRI showed a hemivertebra between T6 and T7 level on the right side with no bearing rib along with secondary right-sided convex thoracic scoliosis and focal thoracic kyphosis at the site of the hemivertebra. His Cobb's angle was 32 degrees preoperatively and he had neither history nor signs of pulmonary hypertension.

Preoperative ECHO showed a patent Glenn shunt, large unrestrictive atrial communications, and good ventricular systolic function. In Figure 1, the patient's hemoglobin level was 16.3 g/dL and his oxygen saturation was 89%. Intraoperatively, standard ASA monitors were attached along with two large-bore IV lines, a radial arterial line, and two central venous lines (CVL). One 5.5 French triple lumen CVL was inserted in the femoral vein to reflect the common atrial pressure (CVP), whereas the other was a 4 French 5 cm double-lumen CVL inserted in the right internal jugular vein, which reflects the pulmonary artery pressure (PAP). Our goal was to keep the transpulmonary gradient (the difference between PAP (Glenn pressure) and CVP) at 3–5 mmHg. In the case of low blood pressure, we increase the Glenn pressure through IV fluids up to 18 mmHg, trying to maintain transpulmonary gradient, and if BP did not improve, ventricular dysfunction and the need for inotropic support are suspected.

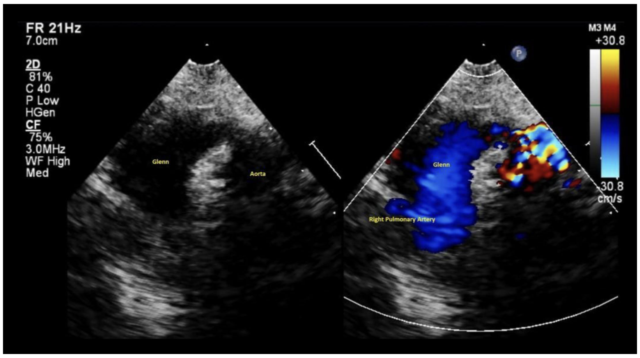

Fig. 1: Glenn echocardiography

IV induction was started using 25 mcg of fentanyl, 25 mg of ketamine, 2 mg of midazolam, and 3 mg of cisatracurium. Intubation was easy. The patient was placed in a prone position. Anesthesia was maintained by total intravenous infusion of propofol and remifentanil. During surgery, his mean arterial pressure (MAP) was maintained above 60 mmHg, his oxygen saturation was in the range of 79%–81%, and his transpulmonary gradient was maintained at 3–5 mmHg. At the end of the operation, the patient was extubated, the central line in the internal jugular vein was removed, and he was sent to the pediatric ICU in a hemodynamically stable condition.

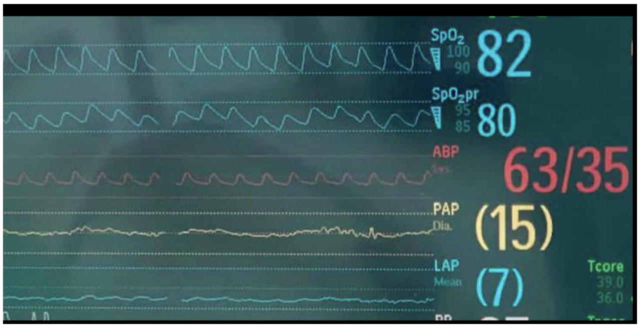

Fig. 2: PAP is the Glenn pressure and LAP is the common atrial pressure

3. Discussion

Scoliosis may produce significant restrictive lung disease.[8] The most important prognostic factor is Cobb's angle, and, respiratory problems are expected with an angle larger than 60 degrees. Patients who have a Glenn shunt have half of their venous return flowing passively from superior vena cava to the pulmonary circulation.[9] The single ventricle is partially unloaded, less stressed, and more resilient and can tolerate most of the major procedures.[6] The anesthetic goals in such a situation are maintaining adequate preload and oxygen delivery by keeping hemoglobin at a higher level, maintaining normal sinus rhythm and adequate ventricular contractility, and attenuating sympathetic stress response to decrease pulmonary vascular resistance.[6] The patient's oxygen saturation baseline was 75%–80%. It is prudent to maintain oxygen content and delivery through ventilation and transfusion, keeping his MAP above 60 mmHg.[2,6,10] In Glenn shunt, we have to keep the Glenn pressure higher than the CVP to keep the circulation flowing.[11,12] When oxygen saturation is lower than 70%, we promote the flow to the lungs by maintaining the transpulmonary gradient by decreasing the pulmonary vascular resistance, which is achieved by increasing fractional oxygen concentration, hyperventilation, deep anesthesia, and avoiding any cause of increased airway pressure.[2] In our case, we inserted two central lines: one was inserted in the right internal jugular vein, with pressures recorded from this line representing the PAP; the other line was inserted in the femoral line, representing the CVP. Our target is to keep a pressure gradient of 3–5 mmHg between the two lines; this will guide fluid management and the need for inotrope in the case of adequate transpulmonary gradient and low blood pressure (Figure 2). In one case report, a 15-year-old boy who had a single left ventricle post-Fontan circulation physiology and idiopathic scoliosis underwent two surgical correction surgeries for his scoliosis due to its high Cobb angle. They achieved hemodynamic stability with a CVP of 20 mmHg.[13]

4. Source of Funding

None.

5. Conflict of Interest

None.

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