Case Report

Anaesthetic management of an adolescent for scoliosis surgery with a bidirectional glenn circulation

Abstract

The relationship between congenital heart disease and scoliosis is well known. Most of these patients present deformities that can be included in the group on spine deformities associated to a congenital heart disease, and, in another patients, the fact of perform a sternotomy in the first infancy to correct partially the heart defect, predisposes to develop the spine deformity. The improvement of the life expectancy in these patients can increase the probability of being operated of non-cardiac surgery. The patients with single ventricle circulation do not have right ventricle, so that, the pulmonary blood flow depends of the central venous pressure. The prone position used for scoliosis correction could worsen the venous return and the cardiac output. We present a 17 years of female patient case, diagnosed of a kippel Feil syndrome and a complex congenital heart disease (dextrocardia, dextroapex, complete atrioventricular septal defect, double outlet right ventricle, transposition of great arteries, pulmonary stenosis and double inferior vena cava system) partially corrected by Glenn surgery at 2 years. She present a toracolumbar scoliosis that causes severe pulmonary restriction and gets worse her functional class. The patient was operated in prone position by posterior approach and the correction using pedicular screws and bars was developed. Hemodynamic invasive monitoring like Transesophageal echocardiogram (TEE) and continuous arterial pressure monitoring were used; somatosensorial evoked potentials were used too. The patient was carried to the intensive care unit hemodinamically stable. As a severe postoperative complication at the second postoperative day she present superior cava vein thrombosis that was not extended to the pulmonary artery anastomosis and that not compromise the Glenn function. She was anticoagulated with unfractionated heparin being the vein successfully recanalized. 10 days after she was discharged to the hospitalization area.

Keywords: congenital heart disease, scoliosis, glenn surgery, superior cava vein thrombosis

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Abbreviations: ICU, intensive care unit; TTE, transthoracic echocardiography; TEE, transesophageal echocardiogram; HR, heart rate; BP, blood pressure; O_2 Sat, hemoglobin oxygen saturation; EFSV, ejection fraction of the single ventricle

Introduction

The association between congenital heart disease and scoliosis is well known. Spine deformities are more common in patients who have been operated of the cardiac disorder by sternotomy or combined sternotomy and thoracotomy1. Kadhim and cols found that 9,8 % of patients operated of single ventricle by Fontan surgery presented radiological evidence of scoliosis 1,1 years after surgery,[1] being the female patients more affected than male patients, showing a ratio of F5: M3. Conversely, Reckles found the same scoliosis incidence in a similar group of patients.[2] The Fontan connection creates a critical upstream and downstream congestion; these features are the basic cause of the major impairments of this circulation. The ventricle cannot compensate the major flow restriction of the Fontan bottleneck: the suction required to compensate for the barrier effect cannot be generated, specifically not in a deprived heart that cannot control the cardiac output and cannot improve the degree of systemic venous congestion. The major determinant of Fontan outcome is the development of pulmonary hypertension determined by the increasing of pulmonary vessels resistance, in which case the Fontan could derive in a shock state and probably in death.[3] Patients with Fontan circulation could have right to left shunting via a patent Fontan fenestration that can increase the risk of paradoxical systemic embolization, situation much increased in prone position.[4]

Preoperative and description of the case

The following case describe a 17 years old female patient with a history of Kippel Feil syndrome, Von Willebrand disease and a complex congenital heart disease (Dextrocardia, dextroapex, complete atrioventricular septal defect, double outlet right ventricle, transposition of great arteries, pulmonary stenosis and double inferior vena cava system), successfully corrected by the development of Double Glenn Surgery at 9 months of life. Was diagnosed of congenital thoracic scoliosis related to the cardiac surgical correction. The disease was progressing from an initial Cobb angle of 15° to an actual angle of 93°, with a right curve that involves the segment between T3 and T9. The symptoms, being dyspnea the most frequently, progressed too, until rest dyspnea with severe functional limitation (Figure 1). She was followed by the pediatric cardiology once a year which developed 2 diagnostic catheterisms by the worsening of the functional class, evidencing a non-significant stenosis of the atrium pulmonary anastomosis and prescribed acetylsalicylic acid 100 mg/ day. At the time of preoperative evaluation, the patient was hemodinamically stable. O_2 Sat 77%, and lung function tests

compatible with severe pulmonary restriction were recorded. She present too a hepatosplenomegaly with a normal coagulation tests and platelets count. Ascites, lower limbs edema or enteropathy were not found. All the predictors of difficult airway intubation were found. TTE was done found the parameters previously described and normal EFSV (66%). Pulmonary pressures were normal too. Antiplatelet treatment with acetylsalicylic acid 100 mg/ day was retired prior to surgery. The patient was premedicated with oral bromazepam. Vital signs. HR 75 bpm, BP 110/60 mmHg O_2 Sat 77% were evidenced at the entrance of the operating room. Intravenous anesthesia induction with etomidate 18 mg, fentanyl 150 mcg was done. Nasotraqueal intubation was performed with the patient awake using a Fiberoptic bronchoscopy. The maintenance was done with propofol/ remifentanil being de doses adjusted according to hemodynamic response and the biespectral index value between 40-60. O_2 Sat was maintained around 80%. 500 mg of tranexamic acid was administered. Central venous access through the right femoral vein and left radial artery were channeled. TEE probe was introduced and the monitoring was performed in three times: after anesthesia induction, after prone position and during the final correction, showing normal flow for bidirectional Glenn and adequate function of the single ventricle.

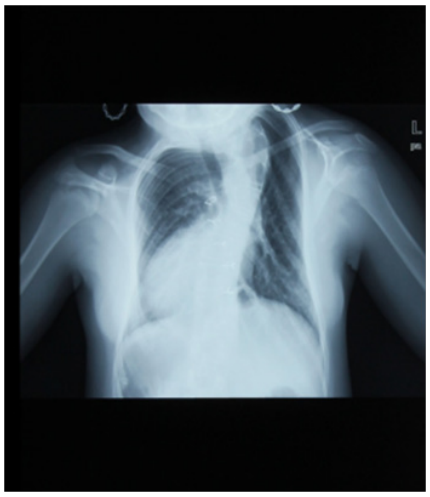

Figure 1 Chest radiography before scoliosis surgery.

The Spine deformity correction was performed by posterior approach placing pedicle screws from T1 to L1 and longitudinal bars to do the correction. Evoked potentials registered were getting worse and for this cause was impossible to correct more than 50% of the scoliosis curve (Figure 2). The hemodynamic parameters were conserved during all the surgery correction except at finally coinciding with a bleeding episode quantified in 800 ml in 3 minutes; presenting hypotension and desaturation (O_2 Sat 69%); In that moment transfusion of 2 packed red cell were required. During the intraoperative period 4000 ml of crystalloid, 1000 ml of colloids and 3 packed red blood cells were intravenous infused. Hemoglobin value of 12.7 at the end of surgery was evidenced and the total urine output registered was 1300 ml. After 9 hours of surgery was carried to the ICU. Three hours later at ICU she presented hypotension and desaturation episode probably related with bleeding, that required additionally transfusion of 2 packed red cells. After this episode she was extubated without incidents. The antiplatelet therapy and the prophylactic low molecular heparin were administered during the first 24 hours of the postoperative period. At the second postoperative day was discharged to the hospitalization area. After 48 hours in this area she presented right cervical edema and facial congestion. Her doctors practiced an TTE that shows superior cava vein thrombosis that involves superior cava vein to the Glenn anastomosis without occluding the pulmonary artery. She was moved back to the ICU and was anticoagulated with non-fractionated heparin, progressing the thrombosis to a total recanalization of the affected vessels 48 hours later. Helicoidally axial tomography was done to discard pulmonary embolism.

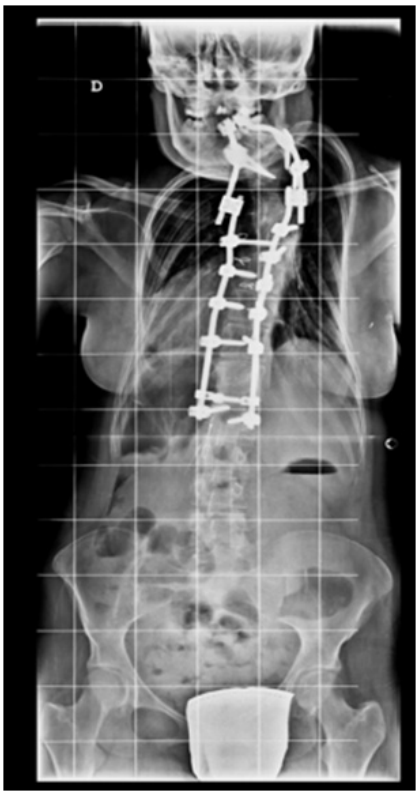

Figure 2 Chest radiography after scoliosis surgical correction.

Discussion

In patients with Glenn physiology, who are going to be operated of non cardiac surgery, the priority should be to maintain an adequate hemodynamic state, for this purpose is important to follow an hemodynamic protocol guided by objectives. Another question is the coagulation impairment induced for the surgery stress, which are of multifactorial etiology, and can trigger a prohemorragic or protrombotic state. For this reason it is necessary to clarify indications of antiplatelet or anticoagulation for such patients to minimize the risk of intraoperative bleeding and to avoid the severe consequences of a thrombosis.Additionally in all patients with single ventricle circulation, is priority to consider that the prone position can worse the venous return and reduce the cardiac output. For this reason preferable these patients must be operated in the Cottrell or in the Jackson table, in which the patient is fastened and the abdomen, and inferior cava vein system, is not compressed.[5] The postoperative complications are frequent being worst the respiratory complications considering that almost all these children are diagnosed of restrictive disorders furthermore their heart disease.[6]

Acknowledgements

None.

Conflicts of interest

The authors declare that there is no conflict of interest.

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References

1. Kadhim M, Pizarro C, Holmes L Jr, et al. Prevalence of scoliosis in patients with Fontan circulation. *Arch Dis Child*. 2013;98(3): 170–175.
2. Reckles LN, Peterson HA, Weidman WH, et al. The association of scoliosis and congenital heart defects. *J Bone Joint Surg Am*. 1975;57(4): 449–445.
3. Gewillig M, Brown SC. The Fontan circulation after 45 years: update in physiology. *Heart*. 2016;102(14):1081–1086.
4. Schwartz MC, Nykanen D, Fleishman C, et al. Fontan Fenestration Closure Prior to Posterior Spinal Fusion in Patients With Single-Ventricle Heart Disease. *Spine*. 2016;41(23):1425–1428.
5. Kadhim M, Spurrier E, Thacker D, et al. Scoliosis surgery in children with congenital heart disease. *Spine*. 2014;39(3): E211–E218.
6. Pérez-Caballero Macarrón C, Sobrino Ruiz E, Burgos Flores J, et al. Spinal surgery in the univentricular heart--is it viable?. *Cardiol Young*. 2014;24(1):73–78.

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