Cyanotic Congenital Cardiac Disease and Scoliosis: Pre-Operative Assessment, Surgical Treatment, and Outcomes

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Significance of the Study

- Children with cyanotic congenital cardiac disease may develop progressive scoliosis requiring surgical correction due to longer life expectancy.
- A multidisciplinary assessment, as well as input from cardiac/haematology colleagues, is mandatory in the peri-operative management and to weigh the risks and benefits of surgery.
- Meticulous surgical technique, hypotensive anaesthesia, tranexamic acid and cell salvage can reduce surgical morbidity.

Keywords

Cardiopulmonary physiology · Cardiovascular disease ·
Scoliosis · Spinal deformity and instability · Spinal surgery

Abstract

Objective: Congenital heart disease (CHD) is associated with the development of scoliosis. Improvements in cardiac care have extended survival of children with cyanotic CHD which possess a need for correction of scoliosis. There is limited information on spinal care for these patients. We present 3 patients with CHD who underwent surgical correction of scoliosis. **Materials and Methods:** We reviewed demographic and clinical data on patients with cyanotic CHD. **Results:** Patient 1 underwent posterior spinal fusion T3–L3 at the age of 16 years. He had a double inlet left ventricle and was treated with completion of a Fontan circulation. Hypotensive anaesthesia was used but he lost 3,000 mL of blood. The operative time was 370 min and most of the blood loss occurred in the

second half of the procedure. Patient 2 underwent posterior spinal fusion T5–T12 when aged 14 years. She had transposition of the great vessels corrected over multiple surgeries. Hypotensive anaesthesia was used, she had blood loss of 300 mL, and the surgical time was 282 min. Patient 3 underwent posterior spinal fusion extending from T5–T12 when he was 17 years old. He had a double inlet left ventricle and was treated with completion of a Fontan circulation. Hypotensive anaesthesia was used, he had blood loss of 1,021 mL, and a surgical time of 342 min. **Conclusion:** Scoliosis surgery in patients with complex cardiac disease may be indicated to treat progressive deformities which produce severe symptoms. A multidisciplinary approach including a spinal surgeon as well as a cardiologist, haematologist, respiratory paediatrician, and spinal anaesthetist can evaluate the general medical condition and weigh the benefits and risks of surgery. Deformity correction can be performed using a meticulous technique and has produced a series of satisfactory outcomes.

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Introduction

Congenital heart disease (CHD) affects 1% of live births and has a recognised association with scoliosis [1]. Current evidence suggests a 10-fold increase in the prevalence of scoliosis in children with CHD when compared to age-matched populations [2]. Contributory mechanisms include congenital vertebral anomalies, thoracogenic causes and the presence of syndromic conditions. Both sternotomy and thoracotomy are associated with the development of scoliosis; children undergoing both procedures do not have a cumulative risk [3–6].

CHD can be separated into cyanotic (10%) and non-cyanotic (90%) types. Cyanotic cardiac disease is a risk factor for major complications following corrective surgery for scoliosis [7, 8]. Amongst the most challenging of cyanotic heart conditions are those with a single ventricular circulation, which includes variations of the single left ventricle (e.g., tricuspid atresia or double inlet left ventricle with or without transposition of the great arteries, malaligned atrioseptal canal with hypoplastic left ventricle) or single right ventricle (e.g., hypoplastic left heart syndrome, double outlet right ventricle with mitral atresia, and heterotaxy syndromes). “Single” ventricle conditions are generally treated in a staged fashion with initial palliation followed at a later stage by completion of a Fontan circulation.

Improvements in the care of patients with cyanotic heart disease have increased survival [9, 10]. This has been driven by a better understanding of the condition, technical advances in surgery (such as the lateral tunnel and extracardiac conduit) and improved technologies (including modified ultrafiltration in cardiopulmonary bypass) [9, 11, 12]. The 25-year survival rate in patients undergoing atriopulmonary connection surgery is reported to be about 75%, while patients undergoing a more modern technique have an expected survival rate of 95% [11]. Given the improved life-expectancy and clear association of congenital cardiac disease with scoliosis, it is expected that more of these patients will present for correction of their scoliosis.

Fontan Circulation

The normal human circulation consists of two connected circuits each having its own “pump.” The pulmonary circulation is powered by the right side of the heart while the systemic circulation receives its blood from the left heart. In single ventricle conditions this pattern is interrupted, resulting in one ventricle powering both systems in tandem, resulting in blood being pumped simultaneously into both the pulmonary and systemic circula-

tions, with the systemic venous return mixing with returning oxygenated blood from the pulmonary circuit. As a consequence, blood pumped into the systemic circulation is a mix of oxygenated and de-oxygenated blood leading to cyanosis with either large amounts of blood to the pulmonary circulation and a reduced volume reaching the systemic circulation, or lower amounts of blood being oxygenated before being mixed with the de-oxygenated systemic venous return and then returned to the systemic circuit. In some rare conditions, the volumes of blood pumped into the pulmonary circulation matches that being pumped to the systemic circulation, resulting in a non-cyanotic patient.

Neonates with a single ventricle usually undergo palliative procedures to restore uninterrupted systemic flow, limit pulmonary blood volume, provide unobstructed pulmonary venous return and inter-atrial flow, as well as to limit atrioventricular valvular regurgitation. Following initial surgery, children undergo additional procedures when older to establish a cavopulmonary circulation. The first stage is to perform a superior cavopulmonary anastomosis which diverts blood from the superior venous circulation (head/upper body) directly into the pulmonary vasculature and harnessing the postcapillary energy to drive the blood through the pulmonary circulation. The other flow to the pulmonary circulation is interrupted so that the single ventricle only pumps blood to the systemic circulation. A significant proportion of the venous return is redirected through the pulmonary circulation where it is oxygenated before returning to the single ventricle. A small amount of cyanosis persists as unoxygenated venous blood from the lower body is returned to the single ventricle and mixes with oxygenated blood from the lungs, and this is especially evident during exercise. A third stage procedure is subsequently performed to connect the lower vena cava to the pulmonary circulation. Prior to 1985, this was done with an atriopulmonary connection, but modern techniques such as the lateral tunnel and the extracardiac conduit have prolonged life expectancy [11]. Given that the pulmonary circulation is driven passively through venous return, these patients have a higher systemic venous pressure, which must be taken into consideration during anaesthesia.

The incidence of congenital cardiac disease is around 1% and only a small proportion of these patients have cyanosis. Previous case reports are limited by variations in underlying diagnosis, differences in patient age undergoing surgery (and hence whether a Fontan circulation has been completed) and the lack of uniformity in the procedures performed. Table 1 summarises all previous

Table 1. Patients with complex CHD treated for spinal deformity

Authors	Patients	Complications	Surgical considerations	Surgical outcome
Leichtle et al. [10]	2	Patient 1: VF during wake-up test requiring CPR	Pre-operative halo traction 2 PSF Mean surgical time: 4.75 h Mean BL: 0.7 L	Patient 1: TH and L scoliosis stabilised Patient 2: TH scoliosis corrected from 120 to 60°, L scoliosis corrected from 62 to 30°
Rafique et al. [13]	1	High intraoperative BL (3.5 L)	PSF	Long-term outcomes not reported
Evans et al. [14]	6 (4 Fontan)	Higher BL in the Fontan group (the authors recommended spinal surgery during the cavopulmonary shunt phase to reduce haemodynamic complications) 1 patient needed bolus adrenaline	2 PSF, 3 GR, 1 growth arrest Mean age: 14.75 years Mean surgical time: 314 min Mean BL: 1.6 L	Mean scoliosis correction 24.2% 1 patient died of cardiac complications 24 months after scoliosis surgery
Kadhim et al. [7]	21 (6 Fontan, 2 univentricular physiology)	2 patients had intraoperative BL > BV 3 patients developed pleural effusions	16 PSF, 5 GR, 2 VEPTR Mean age: 11.1 years Mean surgical time: 5.6 h Mean BL: 1.685 L (PSF)	Long-term outcomes not reported
Pérez-Caballero et al. [8]	8	3 patients needed inotropic support (dopamine) 2 patients developed pleural effusions 1 patient developed pseudarthrosis 1 patient developed wound haematoma	8 PSF Mean age: 14.8 years Mean surgical time: 5.2 h Mean BL: 1.52 L	1 patient died of hypovolaemic shock immediately postoperatively
Hedequist et al. [15]	7	6 patients: 1 Horner syndrome, 1 acute renal tubular necrosis, 2 SMA syndrome, 1 pleural effusion, 1 pleural oedema, 1 UTI, 1 delayed paralysis (48 h postsurgery requiring instrumentation removal and resulting in recovery)	5 PSF, 2 A/PSF Mean age: 14.8 years Mean surgical time: 5.5 h Mean BL: 2.942 L	Long-term outcomes not reported

VF, ventricular fibrillation; CPR, cardiorespiratory resuscitation; PSF, posterior spinal fusion; BL, blood loss; TH, thoracic; L, lumbar; GR, growing rods; BV, blood volume; VEPTR, vertical expandable prosthetic titanium rib; SMA, superior mesenteric artery; UTI, urinary tract infection; A/PSF, anterior/posterior spinal fusion.

case reports or small case series of patients with complex CHD who underwent scoliosis surgery. These studies highlight the high rate of major complications associated with spinal surgery in this group and suggest that a thorough pre-operative evaluation can reduce intra-operative risks and minimise perioperative mortality.

We present 3 patients with cyanotic heart disease who underwent posterior scoliosis surgery in adolescence and were followed beyond skeletal maturity. Two patients had Fontan circulation and 1 had transposition of the great arteries.

Case Presentations

The intra-operative data of our patients is presented in Table 2. Patient 1 presented at age 15 years with a 1-year history of worsening back asymmetry and reported bilateral anterior thigh weakness. He had congenital cyanotic heart disease with double inlet left ventricle, coarctation of the aorta and transposition of the great

arteries that had been treated with pulmonary banding, closure of distal main pulmonary artery with the creation of atriopulmonary window, atrial septectomy, right pulmonary artery stenting and a bidirectional Glenn shunt. He developed a primary right thoracic scoliosis (T6–L1) measuring 66° with secondary left upper thoracic and left lumbar curves. Neurological examination and spinal MRI were normal. Haemoglobin was measured at 205 g/L with a haematocrit of 0.59. Fibrinogen was 2.3 g/L and prothrombin time (PT) was 14 s (INR 1.1). Spirometry showed mild restrictive lung disease with a forced expiratory volume (FEV₁) 77% of predicted, forced vital capacity (FVC) 78% of predicted, and FEV₁/FVC 99% of predicted.

This patient underwent scoliosis correction when he was 16 years old, with the curves measuring 41, 90, and 51°, respectively, through a posterior spinal fusion T3–L3 using pedicle screw/rod instrumentation supplemented by locally harvested autologous and allograft bone (Fig. 1). Aspirin was stopped 2 weeks prior to surgery. The surgical time was 370 min and blood loss was 3,000 mL (106% blood volume). Tranexamic acid (3 g) was administered to prevent excessive blood loss, with a maintenance infusion of 400 mg/h (10 mg/kg/h) throughout the procedure. Blood (750 mL) was

Table 2. Intra-operative patient data

Patient No.	Body weight, kg/ estimated blood volume, mL	Levels of fusion	Duration of surgery, min	MAP, mm Hg	Blood loss, mL (% total blood volume)	Blood products transfused, mL/kg	Clear fluids transfused, mL/kg
1	40.3/2,830	T3–L3	370	56	3,000 (106)	1,380/34.5	5,000/124
2	43/2,800	T5–T12	282	62	300 (11)	303/7.0	1,000/23.3
3	56/4,095	T5–T12	342	68	1,021 (25)	650/11.6	2,600/46

MAP, mean arterial pressure.

collected in a cell saver and transfused. In addition, he received 1 unit of packed red cells, 2 units of fresh frozen plasma (FFP), 1,500 mL of colloid and 3,500 mL of normal saline. Despite his univentricular circulation, hypotensive anaesthesia was pursued with mean arterial pressure (MAP) 56 mm Hg throughout surgery.

After the procedure, he was transferred to the paediatric intensive care unit (PICU) and was extubated 6 h later. High filling pressures were maintained with blood products and crystalloids and no inotropic support was needed. Two additional units of red blood cells and 1 unit of FFP were transfused on the second and third postoperative day. Wound healing was delayed due to localised haemorrhous leakage, which may have been related to the use of low molecular weight heparin (LMWH) immediately after surgery as recommended by our cardiologist. Vitamin K was administered during the first 3 postoperative days. He received nutritional support through nasogastric feeds until the 4th postsurgical day. He spent 4 days in the PICU and was discharged from the hospital on postoperative day 7. Prophylactic aspirin was restarted at discharge. At the latest follow-up (6 years after scoliosis surgery; Fig. 1) this patient had recovered well and radiographs demonstrated correction of the upper thoracic scoliosis from 41 to 17° (59% correction), low thoracic scoliosis from 90 to 19° (79% correction), and lumbar scoliosis from 51 to 8° (84% correction). Spirometry at follow-up showed an improved FEV₁ 85% of predicted, FVC 85% of predicted, and FEV₁/FVC 100% of predicted.

Patient 2 presented at age 13 years with a 1-year history of thoracolumbar pain that required regular analgesia. She had cyanotic cardiac disease with transposition of the great vessels requiring multiple surgeries (atrial septostomy, arterial switching, aortic valvuloplasty, mitral and aortic valve replacement) during childhood. She developed a primary left thoracic (T7–T12) and compensatory right lumbar (L1–L3) scoliosis measuring 48 and 33°, respectively. Neurological examination and pre-operative spinal MRI showed no anomalies. Blood tests revealed a haemoglobin level of 120 g/L and a haematocrit of 0.381. Fibrinogen was 2.7 g/L and PT was 12 s (INR 1.0). Spirometry showed moderate restrictive lung disease with FEV₁ 56% of predicted, FVC 61% of predicted, and FEV₁/FVC 92% of predicted.

She underwent a posterior spinal fusion T5–T12 with the use of pedicle screw/rod instrumentation supplemented by locally harvested autologous and allograft bone when aged 14 years with the scoliosis measuring 58 and 45°. Warfarin was discontinued 5 days pre-operatively. Treatment with LMWH for thromboprophylaxis was stopped the night before surgery. Surgical time was 282 min with a blood loss of 300 mL (11% blood volume). Tranexamic acid (3 g) was administered at induction and main-

tained by infusion (10 mg/kg/h) throughout the procedure. There was insufficient blood captured in the cell saver to allow for transfusion, which resulted in fluid administration with 1,000 mL crystalloids and 1 unit of red blood cells (303 mL). MAP was maintained during surgery at 62 mm Hg.

She was transferred to the PICU extubated and her postoperative course was characterised by intermittent oxygen desaturations with a need for low flow oxygen therapy. A chest X-ray revealed a mild collapse of the left lung which was managed with intense physiotherapy, but she did not require non-invasive ventilation. She received treatment with LMWH during the first 3 postoperative days, after which warfarin was re-introduced. Oral nutrition was gradually established with no need for nutritional supplementation. She spent 3 days in the PICU and was then discharged on postoperative day 6. At the latest follow-up (6 years after surgery) our patient had no clinical symptoms and her scoliosis was corrected to 33° (45% correction) for the thoracic and 24° (47% spontaneous correction) for the lumbar curve. Spirometry at follow-up showed improved FEV₁ 63% of predicted, FVC 68% of predicted, and FEV₁/FVC 93% of predicted.

Patient 3 was 14 years old when he presented with shoulder and waistline asymmetry. His medical history included severe asthma and cyanotic cardiac disease, with double inlet left ventricle and coarctation of the aorta. He underwent multiple cardiac procedures, including pulmonary artery band, Damus bidirectional Glenn, total cavopulmonary circulation completion, and helix device closure of fenestration. His medications included montelukast (leukotriene receptor antagonist), beclomethasone (corticosteroid) and warfarin. He developed a right thoracic scoliosis (T5–T11) measuring 37°. Neurological examination and MRI of the spine showed no abnormalities. The patient was skeletally immature and was kept under observation for the next 3 years when the curve gradually progressed to 55° and a compensatory lumbar curve (L1–L4) developed measuring 27° (Fig. 2). Blood tests revealed haemoglobin levels of 169 g/L with a haematocrit of 0.484 and fibrinogen level of 2.4 g/L. Warfarin was stopped 4 days prior to surgery. Despite discontinuing warfarin treatment 4 days prior to surgery, his PT remained elevated at 19 s (INR 1.7) but the decision was made to proceed with surgery. He was given LMWH for thromboprophylaxis until the night before surgery. Spirometry showed a mixed restrictive/obstructive pulmonary pattern with FEV₁ 60% of predicted, FVC 69% of predicted, and FEV₁/FVC 86% of predicted.

Our patient underwent a posterior spinal fusion extending from T5–T12 using pedicle screw/rod instrumentation supplemented by locally harvested autologous and allograft bone when

Fig. 1. Pre-operative (a, b) radiographs showing a severe triple thoracic and lumbar scoliosis with normal sagittal balance of the spine (patient 1). A posterior spinal fusion produced satisfactory correction of the coronal deformity and a balanced spine in both planes (c, d).

aged 17 years. The surgical time was 342 min and blood loss 1,021 mL (25% blood volume). The cell saver retrieved 650 mL of blood which was transfused. Additional fluid support included 2,100 mL of crystalloids and 500 mL of colloids. The MAP was maintained at 68 mm Hg during surgery. Tranexamic acid was not used.

The patient was transferred to the PICU after surgery extubated but required non-invasive ventilation for 6 h. Anticoagulation was with LMWH and warfarin, which were both given on the first postoperative day. No transfusion was required after surgery.

LMWH was stopped when INR was normal. Oral feedings were gradually initiated. His recovery was uncomplicated. He spent 3 days in the PICU and was discharged from hospital on postoperative day 7. At the latest follow-up (5 years after surgery), he had no problems with his back and radiographs showed the residual thoracic and lumbar curves to measure 20° (64% correction) and 15° (45% spontaneous correction), respectively (Fig. 2). Spirometry at follow-up showed improved FEV₁ 70% of predicted, FVC 75% of predicted, and FEV₁/FVC 93% of predicted.

Fig. 2. Pre-operative (a, b) radiographs showing a primary right thoracic and compensatory left lumbar scoliosis (patient 3). The patient underwent a selective thoracic posterior spinal fusion which corrected the deformity across both curves and produced a balanced effect in both planes (c, d).

Discussion

Spinal deformity surgeons involved in the care of patients with complex CHD should understand the pathophysiology of the condition, as well as the effects of corrective surgery in the heart. It is necessary to preserve high venous pressures in a Fontan circulation and, therefore, hypotensive anaesthesia to reduce intra-operative

bleeding may be less desirable. In addition, these patients are at risk of arrhythmias and may require a pacemaker which can affect the timing and cause possible delay in surgery. Fontan patients may be either hypocoagulable from depletion of clotting factors associated with a protein-losing enteropathy or, more commonly, hypercoagulable due to deficiencies in protein C, protein S, and antithrombin III [9, 11]. Adequate treatment with antico-

agulants requires close collaboration between the cardiologist and haematologist. Whilst many patients who undergo surgery to create a Fontan circulation lead a normal life, it is important to appreciate that while these patients have good circulatory function, their exercise tolerance is reduced, and this can delay their recovery and limit their functional outcomes. It is generally agreed that scoliosis surgery in patients with severe CHD can be beneficial to preserve cardiopulmonary function and prevent progressive cor pulmonale [1, 11].

There is limited information regarding the optimum timing of scoliosis surgery in this group of patients. The 3 patients we treated had evidence of an evolving scoliosis from a young age, as shown by consecutive chest radiographs. Bracing was not attempted on any patient due to the severity of scoliosis and the presence of restrictive lung disease. Others have advocated that spinal surgery be performed at a young age before completion of the Fontan circulation so as to reduce intra-operative blood loss [14]. As our patients were older at the time of surgery, they had larger blood volumes, allowing them to tolerate moderate intra-operative blood loss with no significant consequences. We used controlled hypotensive anaesthesia to reduce surgical bleeding. The patient with the most severe scoliosis required a longer fusion and a prolonged surgical time, resulting in a higher blood loss. Excessive blood loss during the later stages of surgery due to time dependant progressive coagulopathy (intraoperative measurements indicated a platelet count of 113,000 per μL of blood and PT 17 s with INR 1.6) could explain our findings. Surgery in our 3 teenage patients resulted in favourable radiographic correction of their scoliosis which was maintained at follow-up. In addition, their respiratory function, as evident at the most recent spirometry, improved compared to the preoperative state. No patient had back pain at follow-up. Their cardiac function was stable and all patients attended university studies with no

limitations in regards to their back. They reported a better level of function including sports compared to before scoliosis surgery.

Conclusion

Scoliosis surgery for patients with CHD is indicated for those with a progressive, severe deformity. The general health of these patients should be considered and a multidisciplinary approach needs to be used to assess their overall fitness for major surgery. Input from a cardiologist and haematologist is critical in pre-operative tests such as trans-oesophageal echocardiogram and dobutamine cardiac stress testing under general anaesthesia. A detailed coagulation screen should reveal underlying disorders and would guide optimisation strategies to limit peri-operative risks. Close co-ordination of care with the anaesthetists will guide actions, such as the use of hypotensive anaesthesia, tranexamic acid and cell salvage during the procedure. PICU monitoring after surgery is essential in limiting postoperative complications. Our experience suggests that surgical treatment of scoliosis can be performed in this group of highly complex patients, using a meticulous technique with adequate medical support, which can produce satisfactory outcomes that continue into adulthood.

Statement of Ethics

Ethical approval for this study was obtained by the Hospital Research Board.

Disclosure Statement

The authors have nothing to disclose in relation to this study.

References

- Coran DL, Rodgers WB, Keane JF, Hall JE, Emans JB. Spinal fusion in patients with congenital heart disease. Predictors of outcome. *Clin Orthop Relat Res*. 1999 Jul;364:99–107.
- Webb GD, Harrison DA, Connelly MS. Challenges posed by the adult patient with congenital heart disease. *Adv Intern Med*. 1996;41:437–95.
- Herrera-Soto JA, Vander Have KL, Barry-Lane P, Myers JL. Retrospective study on the development of spinal deformities following sternotomy for congenital heart disease. *Spine*. 2007 Aug;32(18):1998–2004.
- Ruiz-Iban MA, Burgos J, Aguado HJ, Diaz-Heredia J, Roger I, Muriel A, et al. Scoliosis after median sternotomy in children with congenital heart disease. *Spine*. 2005 Apr;30(8):E214–8.
- Bal S, Elshershari H, Celiker R, Celiker A. Thoracic sequels after thoracotomies in children with congenital cardiac disease. *Cardiol Young*. 2003 Jun;13(3):264–7.
- Herrera-Soto JA, Vander Have KL, Barry-Lane P, Woo A. Spinal deformity after combined thoracotomy and sternotomy for congenital heart disease. *J Pediatr Orthop*. 2006 Mar-Apr;26(2):211–5.

- 7 Kadhim M, Spurrier E, Thacker D, Pizarro C, Mackenzie WG. Scoliosis surgery in children with congenital heart disease. *Spine*. 2014 Feb;39(3):E211–8.
- 8 Pérez-Caballero Macarrón C, Sobrino Ruiz E, Burgos Flores J, Vázquez Martínez J, Coca Pérez A, Alvarez Rojas E, et al. Spinal surgery in the univentricular heart—is it viable? *Cardiol Young*. 2014 Feb;24(1):73–8.
- 9 Marino BS. Outcomes after the Fontan procedure. *Curr Opin Pediatr*. 2002 Oct;14(5):620–6.
- 10 Leichtle CI, Kumpf M, Gass M, Schmidt E, Niemeyer T. Surgical correction of scoliosis in children with congenital heart failure (Fontan circulation): case report and literature review. *Eur Spine J*. 2008 Sep;17(S2 Suppl 2):S312–7.
- 11 d’Udekem Y, Iyengar AJ, Galati JC, Forsdick V, Weintraub RG, Wheaton GR, et al. Redefining expectations of long-term survival after the Fontan procedure: twenty-five years of follow-up from the entire population of Australia and New Zealand. *Circulation*. 2014 Sep;130(11 Suppl 1):S32–8.
- 12 Gewillig M. The Fontan circulation. *Heart*. 2005 Jun;91(6):839–46.
- 13 Rafique MB, Stuth EA, Tassone JC. Increased blood loss during posterior spinal fusion for idiopathic scoliosis in an adolescent with Fontan physiology. *Paediatr Anaesth*. 2006 Feb;16(2):206–12.
- 14 Evans S, Ramasamy A, Marks DS, Spilsbury J, Miller P, Tatman A, et al. The surgical management of spinal deformity in children with a Fontan circulation: the development of an algorithm for treatment. *Bone Joint J*. 2014 Jan;96-B(1):94–9.
- 15 Hedequist DJ, Emans JB, Hall JE. Operative treatment of scoliosis in patients with a Fontan circulation. *Spine*. 2006 Jan;31(2):202–5.