Surgical correction of scoliosis in children with congenital heart failure (Fontan circulation): case report and literature review

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Received: 13 March 2007 / Revised: 9 December 2007 / Accepted: 22 December 2007 / Published online: 29 January 2008
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Abstract Patients with Fontan circulation represent a significant cardiorespiratory risk during spinal surgery. We report about two patients with severe scoliosis and Fontan circulation and their successful operative treatment. The case report will be compared with the national and international literature.

Keywords Scoliosis · Congenital heart failure · Fontan circulation

Introduction

Patients with cyanotic conditions have a fourfold increased risk of developing scoliosis [3]. The survival-rates in patients with complex congenital heart disease are improving, thus creating a subset of patients with complex physiology who will require surgical treatment of their scoliosis. The reasons for surgical correction are, an increasing imbalance in combination with back pain, possible secondary additional impairment of the cardio respiratory function and lack of conservative treatment options. The aim of the operative therapy is the frontal and

sagittal correction of the deformity with a primary stable instrumentation that enhances a solid spondylodesis and requires no postoperative brace treatment. We report about two patients with cyanotic conditions (Fontan physiology) and severe scoliosis and successful posterior correction and instrumentation.

Case report

During their early childhood both patients underwent a thoracotomy and/or sternotomy and cardiac surgery to facilitate more physiological blood flow (Fontan physiology). In this operation the systemic blood flow of the univentricular heart is diverted directly into the pulmonary artery bypassing the right ventricle by creating a cavo-pulmonary shunt for venous return to go into the pulmonary vasculature with resultant oxygenation. To maintain a continuous blood flow, these patients require a higher systemic venous pressure.

Patient 1 is a 14-year-old girl with a double-curved scoliosis of thoracal 85° and lumbar 70° Cobb angle. Despite the Fontan physiology, she also had a pace maker which inhibited the monitoring of motor-evoked potentials (MEP's) during surgery. A total intravenous anaesthesia (TIVA) was used with a CVP and arterial line and defibrillation pads were placed. We additionally did not use bipolar coagulation, which retrospectively would have not been necessary. After instrumentation from T5-L4 and correction, a wake-up test was performed to check on the neurological function. During this test, the patient developed ventricular fibrillation and cardiopulmonary resuscitation was necessary while the situs was only temporarily closed. After stabilisation of the cardio respiratory circulation, the operation could be finished

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with minor correction of the thoracic curve (60°) and the lumbar curve of 55° (Figs. 1, 2). The time taken for the operation was 360 min, including 2 h of successful resuscitation. The blood loss was 800 ml. The postoperative course was uneventful. The patient was discharged two weeks after surgery. No brace was necessary and a good clinical outcome without loss of correction after 4 years was achieved.

Patient 2 is a 16-year-old girl with a progressive right thoracic scoliosis of 120° and a lumbar curve of 62° (Figs. 3, 4). Because of the severity of the curve and the impaired cardio respiratory function (Fontan physiology),

we began treatment with Halo-extension for 2 weeks pre-operative to reduce the thoracic curve to 80° . Therefore, a Halo-ring was assembled and the primary extension load of 3 kg was daily increased by 1 kg until the half body weight was nearly reached. During surgery again, we used a cardio specific anaesthesia (TIVA) and a trans oesophageal echocardiography (TOE). Additionally, continuous spinal monitoring with SEP's and MEP's was performed, which required no intra operative wake-up test. Posterior pedicle screw instrumentation and correction was performed from Th2 to L3. The time taken for the operation was 210 min with a blood loss of 600 ml. The thoracic curve could be

Fig. 1 Patient 1. Double-curved scoliosis of thoracic 85° and lumbar 70° Cobb angle. **a** ap view, **b** lateral view

Fig. 2 Patient 1. The thoracic curve could be corrected to 60° and the lumbar curve to 55° Cobb angle. **a** ap view, **b** lateral view

corrected to 60° and the lumbar curve to 30°. The post-operative course was uneventful with mobilisation brace-free. The patient was discharged on the 13th postoperative day. A good clinical outcome 12 months postoperative with no loss of correction was achieved.

Discussion

There is an increased incidence of severe spinal deformities in children with congenital heart disease that will require corrective surgery. The deformities are not specifically

related to the cardiac defects and may be congenital, idiopathic or thoracogenic [1]. Because of multiple operations of the heart in the childhood anterior correction is mostly difficult and because of the pulmonary situation, not recommended. With a cavopulmonary shunt, a higher systemic venous pressure is required and intra operative induced hypotension is not possible. In this special group of patients the cardio respiratory morbidity is increased due to the possible development of cardiac dysarrhythmias and an increased risk of thromboembolic events [4].

To our knowledge, there exists only one case-series of seven patients about the surgical correction of spinal

Fig. 3 Patient 2. Right thoracic scoliosis of 120° and a lumbar curve of 62° Cobb angle. **a** pa view, **b** lateral view

deformities in children with Fontan circulation. Hedequist et al. [2] reported about five patients who underwent posterior instrumentation and two patients who underwent anterior and posterior procedures. The mean preoperative curve of 75° could be corrected to 35°. The authors report about a postoperative complication rate of nearly 100%. One patient developed a delayed postoperative motor and sensory palsy. A posterior revision and removal of the instrumentation was necessary. One patient developed a Horner syndrome; a reversible superior mesenteric artery syndrome was seen in two patients. One patient developed a tubular necrosis and two others a pulmonary edema and pleural effusion, respectively, requiring therapy. Hedequist et al. [2], therefore postulate that short operation times without prolonged attempts of instrumentation and correction and minor loss of blood leads to a better clinical outcome. In addition to this, the choice of a suitable anaesthesia in cooperation with an experienced paediatric cardiac anaesthesia team is decisive for successful therapy. In the literature TIVA is recommended as the anaesthetic procedure of choice. Vischoff et al. [4] established recommendations for the anaesthetic management for scoliosis surgery with Fontan circulation. In addition to the basic

monitoring, the central venous pressure (CVP) should be measured and a permanent transoesophageal echocardiography should be used to visualize even a minimal ventricular filling period. To stabilize the circulation the haemoglobin should be kept stable with regular blood transfusions during operation. Moreover, regular blood transfusions are useful to prevent a reduced perfusion of the myelon and other organs (kidney, GIT). In our opinion, this is why a permanent spinal cord monitoring should be preferred to an intraoperative wake-up test. Beyond the measurement of the SEP's and MEP's, a reduced perfusion of the spinal cord can be detected sooner. Because of the surgical trauma and the heart disease, the wake-up test constitutes an increased cardiac loading in these children. Therefore, cardiorespiratory complications with possible serious consequences are at a higher risk. In our two cases Patient 1 developed an unstable cardio respiratory condition during the wake up test that required resuscitation, while the situs was not closed. Fortunately, the girl recovered without consequences.

In summary, evaluation and monitoring of cardio respiratory specific parameters including coagulation and the intra operative management with an experienced pediatric cardiac anaesthesia team is compulsory. In severe curves

Fig. 4 Patient 2. The thoracic curve could be corrected to 60° and the lumbar curve to 30° Cobb angle. **a** pa view, **b** lateral view

Halo-extension allows a good preparation and enhances the surgical correction, which should be performed using a primary stable posterior instrumentation with pedicle screws. While preparing for such operations, an extensive anaesthetic and orthopaedic consent is necessary to prepare the parents for the possible consequences. In our two patients a good clinical and functional cure was achieved (both girls attending school as regular students), which is reflected in a high degree of satisfaction of the patients and their families.

Conflict of interest statement None of the authors has any potential conflict of interest.

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