Case Report

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Hemodynamic deterioration due to increased anterior and posterior cardiac compression during posterior spinal fusion for scoliosis with pectus excavatum

Ryota Adachi[1], Tasuku Nishihara[1], Tadao Morino[2], Keisuke Sekiya[1], Sakiko Kitamura[1], Amane Konishi[1], Yasushi Takasaki[1], Hiromasa Miura[2], Naoki Abe[1] and Toshihiro Yorozuya[1]

Abstract

Hemodynamics may deteriorate during the perioperative period when performing posterior spinal fusion in patients with pectus excavatum and scoliosis. A 13-year-old teenager diagnosed with Marfan syndrome had thoracic scoliosis and pectus excavatum. Thoracic scoliosis was convex to the right, and a right ventricular inflow tract stenosis was observed due to compression induced by the depressed sternum. The patient underwent T3–L4 posterior spinal fusion surgery for scoliosis. Deterioration of hemodynamics was observed when the patient was placed in the prone position or when the thoracic spine was corrected to the left front. Postoperative computed tomography examination showed that the mediastinal space was narrowed due to the corrected thoracic spine. Special attention should be paid in the following cases: (1) severe pectus excavatum, (2) right ventricular inflow tract compression due to depressed sternum on the left side, (3) correction of the thoracic spine on the left front, (4) long-term surgery, and (5) risk of massive bleeding. In some cases, pectus excavatum surgery should be prioritized.

Keywords

Pectus excavatum, scoliosis, Marfan syndrome, posterior spinal fusion, Haller index, right ventricular inflow stenosis

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Introduction

Pectus excavatum is the most common morphological abnormality of the thorax, with an incidence of approximately 1 per 400–1000 live births.[1] Approximately 20% of patients with pectus excavatum have scoliosis.[2,3] In patients with scoliosis and pectus excavatum, the sternal position is displaced from the center of the vertebral body, and therefore, the modified Haller index is used to evaluate the severity of pectus excavatum.[4] The modified Haller index is measured using the vertical distance between the posterior edge of the deepest point of the sternum and the parallel line on the anterior edge of the vertebral body. Scoliosis and pectus excavatum are more common in patients with Marfan syndrome, and their deformities are also marked.[5] The sternum depression located on the left side can directly compress the right ventricular inflow tract anteriorly.[4] The prone position for posterior spinal fusion surgery can increase the anterior cardiac compression and worsen hemodynamics. Furthermore, moving the right-convex thoracic spine to the left front with spinal fusion may exacerbate the posterior cardiac compression due to the corrected spine and cause deterioration of hemodynamics.[6] We report a case of a patient with Marfan syndrome with pectus excavatum and scoliosis who had hypotension and tachycardia during surgery due to the prone position assumed and correction of the thoracic spine.

[1]Department of Anesthesia and Perioperative Medicine, Graduate School of Medicine, Ehime University, Toon, Japan
[2]Department of Orthopaedic Surgery, Graduate School of Medicine, Ehime University, Toon, Japan

Corresponding Author:
Naoki Abe, Department of Anesthesia and Perioperative Medicine, Graduate School of Medicine, Ehime University, Shitsukawa, Toon 791-0295, Ehime, Japan.
Email: abecometen422@yahoo.co.jp

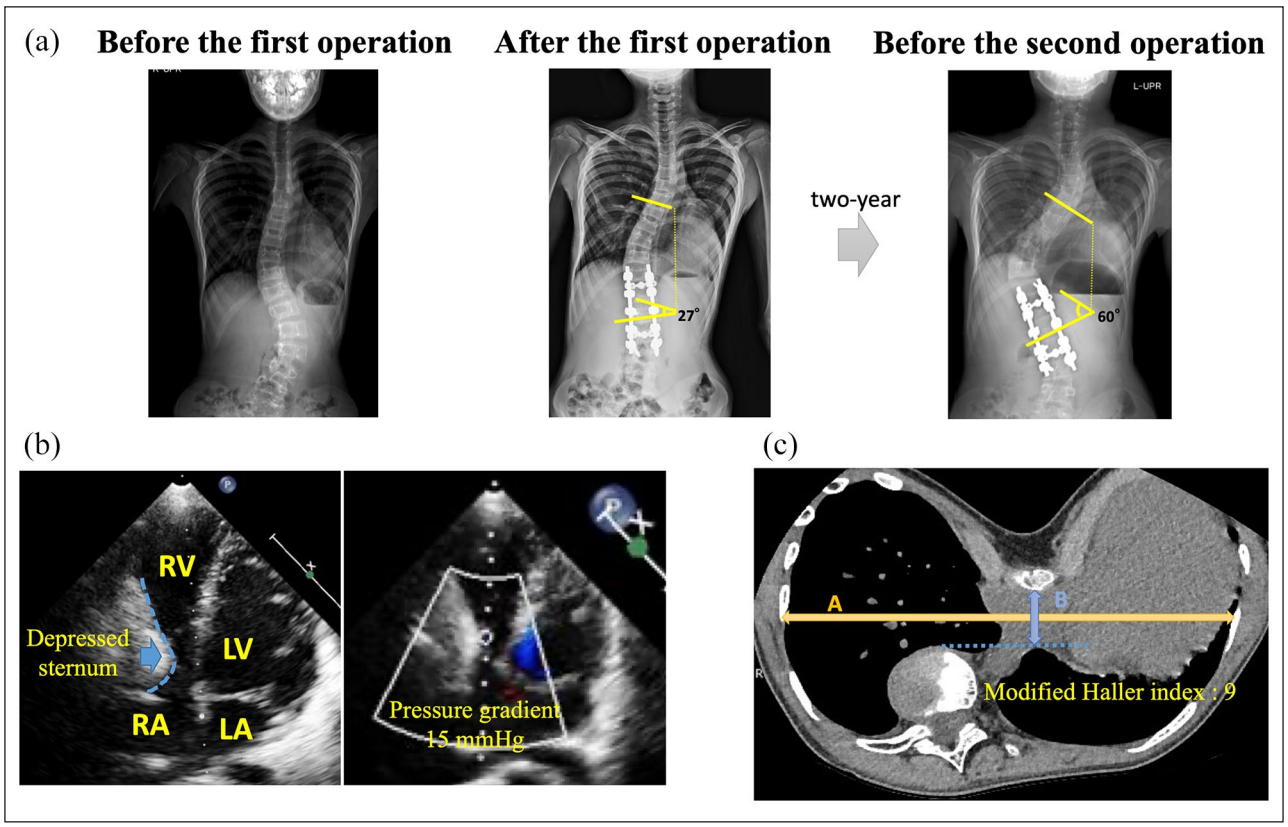

Figure 1. Preoperative examination. Chest X-ray photograph (Xp), transthoracic echocardiography, and chest computed tomography (CT) examination. Over a 2-year period, the Cobb angle deteriorated from 27° to 60° (a). The maximum right ventricular inflow pressure gradient was 15 mmHg (b). The preoperative modified Haller index was 9 (c). The modified Haller index was obtained as the ratio A/B. Values >3.5 are considered to be associated with severe pectus excavatum cases.
LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

Case report

A 13-year-old teenager was scheduled to undergo T3–L4 posterior spinal fusion surgery for thoracic scoliosis. The patient was diagnosed with Marfan syndrome at the age of 11 years and had pectus excavatum. He had no other medical history. Two years before surgery, T12–L4 posterior spinal fusion was performed due to lumbar scoliosis. At that time, there was no particular problem during the perioperative period. Subsequently, as the scoliosis of the thoracic spine progressed (Figure 1(a)), it was decided to operate again. Preoperative evaluation showed that the % vital capacity (% VC) was reduced to 52.3%. Respiratory dysfunction due to scoliosis and thoracic deformity were observed. The maximum right ventricular inflow pressure gradient, which indicates the degree of anterior compression, increased from 12 to 15 mmHg (Figure 1(b)). The modified Haller index was severe but did not change (Figure 1(c)). The thoracic scoliosis was convex to the right side, and the sternal depression was located on the left side (Figure 1(a) and (c)). Anesthesia induction and tracheal intubation were performed without any problems. Immediately after the patient was placed in the prone position, his blood pressure decreased to 74/51 mmHg and the heart rate increased to 134 beats per minute (Figure 2(a)). Because maintaining circulation was possible by adjusting the body position, administering several bolus doses of 50-μg phenylephrine, and loading an infusion solution, we decided to continue the operation. The scoliosis was corrected according to the hybrid method. We bent the titanium rod to the desired degree of thoracic kyphosis and lumbar lordosis. The rod was attached to the hook on the left cranial side (T3,6) and the screw head on the caudal side (T11, 12, L4), and the scoliosis and sagittal alignment were corrected by rotating them. The sublaminar tape passed through T4,5, and 7-10 was attached to the rod to pull and fix the vertebral body. In addition, a rod was installed on the right side. By compressing the convex side and distracting the concave side, further correction was performed and the shoulder balance was adjusted. After the onset of the thoracic spine correction, the heart rate increased and blood pressure decreased gradually despite the autologous blood transfusion and vasopressor administration (Figure 2(b)). At the end of the surgery, the systolic blood pressure decreased to 60 mmHg. After placing the patient in the supine position, the hemodynamics rapidly stabilized (Figure 2(c)). The duration of the operation was 3 h 19 min, the blood volume

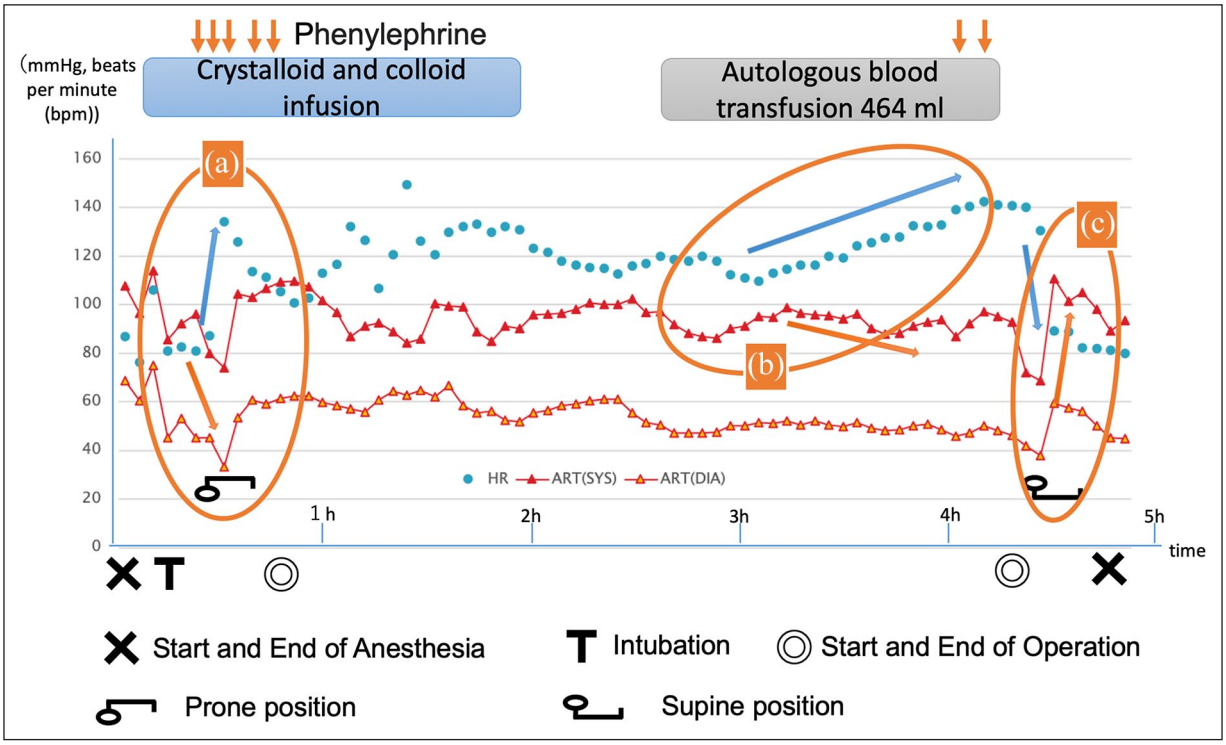

Figure 2. Intraoperative progress. Hypotension and tachycardia due to the patient's prone position (a) and correction of the thoracic spine (b). Stabilization of hemodynamics due to the patient's supine position (c).
ART(SYS): systolic blood pressure, ART(DIA): diastolic blood pressure, bpm: beats per minutes, HR: heart rate.

attributed to bleeding was 790 ml, the infusion volume was 2500 ml, and the blood transfusion volume was 464 ml. Mechanical ventilation was continued in the intensive care unit after the operation. The patient was extubated on the same day. Postoperative imaging showed that the Cobb angle of the thoracic spine was corrected from 60° (T6–L4) to 32° (T7–L1), and the thoracic spine was moved to the left (Figure 3). The thoracic kyphosis changed from 39° to 27°, and the thoracic spine was moved forward (Figure 3). Consequently, the space in the mediastinum decreased, and the cardiac compression due to the corrected thoracic spine and depressed sternum became stronger (Figure 3). The spinal penetration index, which quantifies the portion of the rib cage occupied by vertebrae, increased from 10% to 16%.[7] The modified Haller index increased from 9 to 13.4.

Discussion

Marfan syndrome is an inherited connective tissue disorder with ocular, musculoskeletal, and cardiovascular manifestations that are caused by mutations in fibrillin-1 and transforming growth factor β 1, 2, (TGFβ receptor).[8] Musculoskeletal abnormalities, such as pectus excavatum and scoliosis, constitute some of the major diagnostic criteria for Marfan syndrome.[9] In patients with abnormally soft cartilage, such as those with Marfan syndrome, negative intrathoracic pressure can induce a concavity of the anterior thoracic wall that primarily consists of costal cartilages.[10] Therefore, patients with Marfan syndrome often have severe thoracic deformities. Severe pectus excavatum may compromise respiratory and cardiovascular function because the lungs and heart are mechanically compressed by the depressed sternum.[11,12] In our patient, right ventricular inflow tract stenosis and restrictive ventilatory impairment were also observed. In such cases, it is important to evaluate the degree of pectus excavatum and the positional relationship between the heart, sternum, and thoracic spine by preoperative computed tomography (CT) examination. This is because it is possible to predict the adverse effects of cardiopulmonary function due to prone position and correction of the thoracic spine.

In the prone position of a patient with pectus excavatum, hemodynamics may be disrupted due to increased anterior cardiac compression from the depressed sternum (Figure 4(a)). If the sternal depression is on the left side, the right ventricular inflow tract can be compressed directly. Galas et al. showed that prone positioning of a patient with pectus excavatum exacerbated the right ventricular inflow tract stenosis.[13] One report stated that the Nuss procedure was prioritized over posterior spinal fusion because of a marked decrease in blood pressure due to the patient's prone position.[14] Moreover, another report indicated that posterior spinal fusion progressed uneventfully after anterior compression due to the fact that the prone position could be modified by

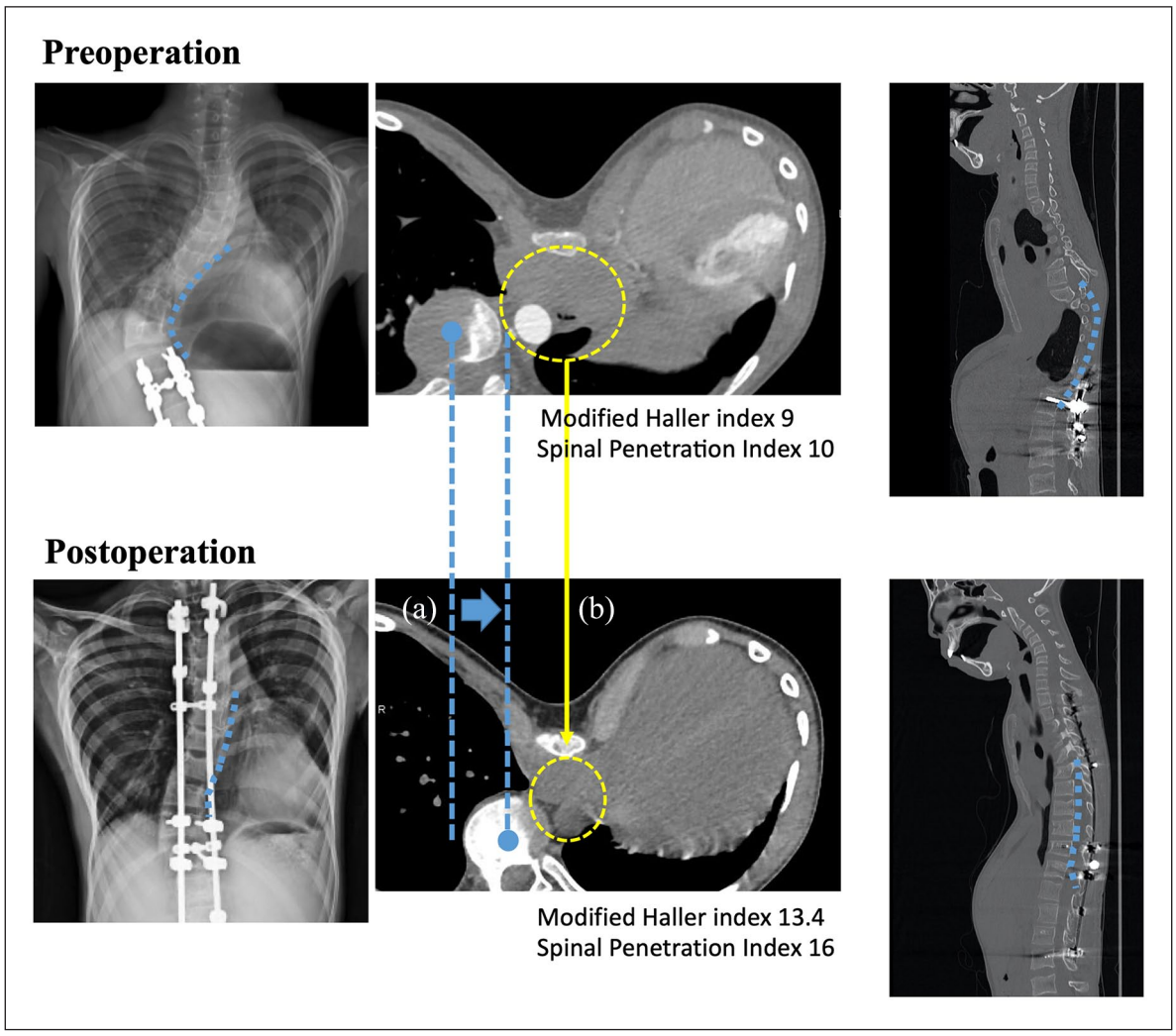

Figure 3. Preoperative and postoperative chest X-ray photograph (Xp) and computed tomography (CT) examinations. Left movement of the thoracic spine (a). Narrowed mediastinum (b). In this case, the modified Haller index increased from 9 to 13.4 and the spinal penetration index increased from 10 to 16.

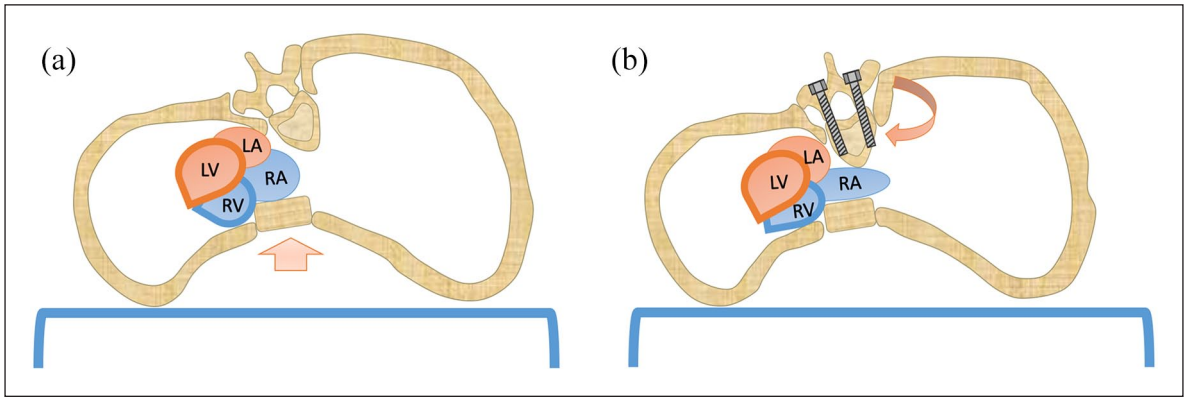

Figure 4. Mechanism of hemodynamic deterioration due to the prone position and spinal correction. Increased anterior cardiac compression due to prone position (a). Increased posterior cardiac compression attributed to the corrected spine (b).
LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.

Table 1. Previously reported cases of serious hemodynamic compromises.

A: Hemodynamic compromises associated with prone position (anterior compression).			
Author	Case	Preoperative evaluation	Details
Galas et al.[13], Bafus BT et al.[14]	Male (15 years old)	Peak right ventricular inflow pressure gradient of 7 mmHg	Immediately after being placed in the prone position, severe hypotension was developed despite volume and pharmacologic treatments. The spinal fusion was aborted. Pectus excavatum repair was conducted prior to scoliosis surgery.
Alexianu D et al.[15]	Boy (34 months old)	Mild compression of the right ventricle	The systolic blood pressure rapidly declined after the patient was positioned prone on transverse bolsters. After the compression of the right ventricle was reduced based on the adjustment of the position using a longitudinal bolsters situated along the side of his body, posterior spinal fusion progressed uneventfully.
B: Hemodynamic compromises associated with spinal corrections (posterior compression).			
Uvodich M et al.[16]	Male (16 years old)	Haller index increased from 8.3 to 11.2 after scoliosis surgery	Hemodynamic instability was noted after the patient was placed in a prone position. Surgical procedure proceeded under vasopressor support and fluid therapy. Hemodynamic instability was continued despite the fact that the patient returned to the supine position. The hemodynamical response of the patient slowly improved without additional surgical intervention.
Lohnhardt M et al.[17]	Male (15 years old)	Subtotal compression of IVC proximal to the right ventricle	During the surgical reduction of the spine for relordosation, a significant drop in blood pressure occurred. Following sufficient resuscitation, the surgical procedures were completed. The patient developed IVC compression symptoms postoperatively. After the Nuss procedure was performed, the symptoms improved significantly.
Rouch A et al.[18]	Girl (14 years old)	Haller index increased after scoliosis surgery Compression of IVC between spine and sternum	No intraoperative complication was noted. Ascites and pleural effusion associated with the compression of IVC were noted postoperatively. After the Ravich intervention was performed, the symptoms improved significantly.

IVC: inferior vena cava.

adjusting the position using longitudinal bolsters.[15] Similar to previously reported cases, in our case, the sternum depression was also located on the left side. Hence, if the sternum compressed the right ventricular inflow tract, the prone position may cause disruption of hemodynamics. These case reports are summarized in Table 1.

In addition, in the case of the correction of right-convex thoracic scoliosis and thoracic kyphosis, the corrected thoracic spine may compress the heart posteriorly (Figure 4(b)). Hypotension caused by increased posterior cardiac compression due to the corrected thoracic spine may persist even after returning to the supine position.[16] In these cases, it is necessary to release the cardiac compression, and an additional pectus excavatum surgery is considered. Lohnhardit et al.[17] and Rouch et al.[18] reported an inferior vena cava (IVC) syndrome case after posterior spinal fusion. The IVC was sandwiched between the depressed sternum and the corrected thoracic spine, and the Haller index increased postoperatively. Additional Nuss or Ravitch procedures improved the symptoms of IVC compression. Tauchi et al. predicted the possibility of these complications in 2018.[4] Their report demonstrated that the Haller index increased postoperatively in approximately 50% of patients. In our case, the right-convex thoracic scoliosis was corrected on the left front side, and the postoperative mediastinal spaces were smaller after the operation. Therefore, the risks of these complications may have been high. In such cases, it is important to plan the surgery considering the possibility that the correction of the thoracic spine could increase the cardiac compression. Table 1 summarizes the cases of complications caused by increased posterior cardiac compression from the corrected thoracic spine.

Cardiac compression may result in venous congestion and may increase the risk of surgical bleeding. In fact, scoliosis surgery in patients with Marfan syndrome tends to have a high-average bleeding volume of $\geqslant$1700 ml.[19] This is because the thorax is soft and the anterior cardiac compression due to the prone position increases. These vulnerable patients can easily experience circulatory collapse due to prolonged surgery and bleeding. Therefore, patients with

Marfan syndrome who have scoliosis and pectus excavatum may often require Nuss surgery for hemodynamic stabilization. However, patients with Marfan syndrome may require emergency surgery due to aortic dissection. In such cases, the Nuss bar can render an emergency surgery difficult. Hence, this disadvantage must be considered when deciding on the indication for Nuss surgery in patients with Marfan syndrome. If Nuss surgery is not performed despite the high risks of circulatory collapse on preoperative assessment, transesophageal echocardiography should be used to monitor the degree of cardiac compression. When circulatory collapse occurs during surgery, we must consider releasing the correction and changing the patient to the supine position during the operation.

Conclusion

Hemodynamics may deteriorate during the perioperative period when posterior spinal fusion is performed in patients with pectus excavatum and scoliosis. Special attention should be paid in the following cases: (1) severe pectus excavatum, (2) right ventricular inflow tract compression due to the depressed sternum located on the left, (3) correction of the thoracic spine to the left front side, (4) long-term surgery, and (5) risk of massive bleeding. In some cases, pectus excavatum surgery should be prioritized. If posterior spinal fusion is prioritized, the degree of anterior cardiac compression caused by the prone position and that of posterior cardiac compression due to spinal correction should be monitored using transesophageal echocardiography.

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Authors' contributions

RA wrote the article draft. TN supervised writing of the article. TM is the operator for this case and edited the article. KS and AK supported data collection and preparing the figures. SK and YT supported perioperative management. HM edited the article. NA supervised anesthesia management and completed the article. TY edited the article. All authors have read and approved the article of this case report.

Availability of data and material

Datasets supporting the conclusions of this article are included in the article.

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from a legally authorized representative (the father of the patient) for anonymized patient information to be published in this article.

ORCID iD

Naoki Abe https://orcid.org/0000-0001-7096-8883

Supplemental material

Supplemental material for this article is available online.

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