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CASE REPORT

Rescue Nuss procedure for inferior vena cava compression syndrome following posterior scoliosis surgery in Marfan syndrome

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Abstract

Purpose Scoliosis surgery in Marfan syndrome is common, even in the presence of a funnel chest. However, to date, no case has been reported with acute intra-/postoperative decompensation caused by vena cava compression following posterior spinal derotation and fusion.

Methods A 15-year-old male patient with Marfan syndrome, a funnel chest and severe scoliosis was treated with surgery for the spinal deformity. Intraoperatively, the patient developed a clinically relevant compression of the inferior vena cava with severe circular depression. Postoperatively, a cava compression syndrome with severe pleural effusion, ascites and enormous swelling of the lower limbs was developed. A conservative treatment of the symptoms, consisting of thoracic drainage and negative fluid balance, failed. Subsequently, the patient was transferred to pediatric intensive care unit for further treatment. Echocardiography and a CT scan demonstrated cava compression syndrome. A rescue Nuss procedure of the funnel chest deformity was performed since conservative treatment failed. The clinical course proceeded without complications and with a decrease in clinical symptoms of inferior inflow congestion. The patient was discharged after almost 3 weeks.

Conclusion The problem of congenital stenosis of the inferior vena cava in Marfan syndrome has not yet been investigated. In the case of simultaneously existing funnel chest and scoliosis in Marfan syndrome, an interdisciplinary discussion is required to decide whether a repair of the funnel chest should be performed first in order to prevent a clinically relevant compression syndrome. For the detection of a preoperatively relevant stenosis of the inferior vena cava, an MRI or thoracic/abdominal CT should be used preoperatively.

Keywords Marfan syndrome · Funnel chest · Posterior spinal fusion · Nuss procedure · Inferior vena cava syndrome · Inferior inflow congestion

Introduction

Scoliosis surgery in Marfan syndrome is common with the presence of a funnel chest (pectus excavatum). However, to date, no case of a postoperative decompensation following posterior spinal correction caused by vena cava compression has been described in the literature. In particular, a consequential Nuss procedure to repair the funnel chest in order to reduce pressure on the vein has not yet been analyzed.

The funnel chest is the most common morphological abnormality of the ventral chest wall, with an incidence of 1–8 per 1000 live births [1]. A high incidence is observed in male patients with Marfan syndrome. It typically results in an asymmetrical pectus excavatum and is used as a diagnostic criterion for Marfan syndrome [2].

The Nuss procedure (according to Nuss, 1998) [3] is a minimally invasive technique that can be described as the gold standard for correcting a funnel chest, due to its good cosmetic results and technical effectiveness. In the past, the procedure has mainly been used in children with symmetrical chest wall configurations; however, with modifications, it is now also used for asymmetrical repairs in adult patients.

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This surgery is considered an elective and should only be carried out after thorough planning and preparation [4, 5]. The presented case study is the first to describe Nuss surgery being performed as a rescue procedure. A posterior revision and reduction of the scoliosis correction had previously been discussed; however, due to the patient's wishes and a resulting of balance syndrome, this avenue was not pursued.

Case

The patient (15/m) was diagnosed with Marfan syndrome at the age of 8, due to scoliosis and an extreme manifestation of a pectus excavatum (Figs. 1, 2). Progressive scoliosis, severe back pain and a loss of spinal mobility after a growth spurt confirmed the indication for the posterior spinal fusion. A preoperative CT scan showed the scoliosis and pectus excavatum with a clinical irrelevant compression of the vena cava inferior (Figs. 3, 4). A dorsal instrumentation from T4 to L4 vertebrae was carried out (Fig. 5). During the surgical reduction of the lumbar spine for relordosation, a significant drop in blood pressure occurred, requiring major fluids. Intraabdominal blood loss appeared to be the cause of this complication. The reposition was ceased, and the circulation

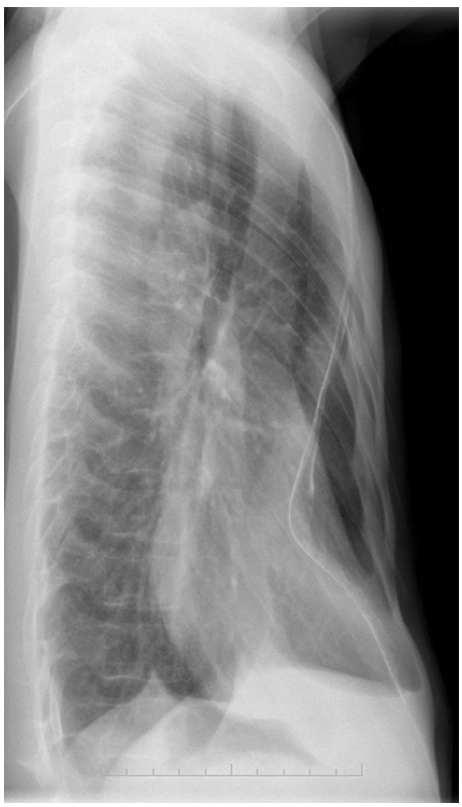

Fig. 1 X-ray showing the thorax preoperatively with obvious signs of a funnel chest

Fig. 2 X-ray showing the spine preoperatively with obvious scoliosis

was stabilized by extensive transfusion of 800 ml cell-saver-blood, 4 red cell concentrates, 1 platelet concentrate, 6 plasmas and 2000IE PCC (prothrombin complex concentrate). Following the transfusions, the dorsal reduction was performed, and the surgical procedures were completed with no further complications. The patient was transferred, intubated and ventilated, to the pediatric intensive care unit.

Following sufficient circulatory resuscitation, the patient was extubated. Due to increasing cardiac distress and a large right pleural effusion, a thoracic drainage was placed with an initial discharge of 1000 ml fluid. On the second postoperative day, the abdomen was distended with ascites, and compressed organs (e.g., liver) and diaphragmatic elevation were detected. Negative fluid balance and substitution of albumin showed a positive effect on the patient's clinical condition, and the thoracic drainage was removed. On the fourth postoperative day, the patient's clinical condition deteriorated rapidly, with large pleural effusion in the right lung and an increasing amount of free intraabdominal fluid. The pleural effusion was again treated with a thoracic drain.

Fig. 3 Preoperatively visible constriction of the inferior vena cava; CT sagittal image

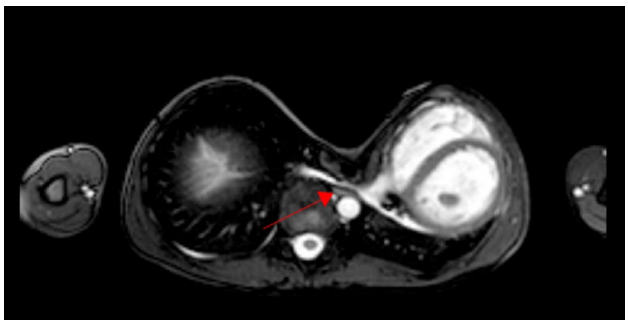

Fig. 4 Preoperatively visible constriction of the inferior vena cava; CT transversal image

The postoperative CT scan of scoliosis surgery suggested a subtotal compression of the vena cava inferior in proximity to the right ventricle, between the sternum and the spine. Furthermore, the scan showed pleural effusions in both lungs and a compression atelectasis of the left basal lung.

Then, all the patients showed severe cardiovascular depression due to severe vena cava compression syndrome. Postoperatively, the patients developed additional symptoms including right pleural effusion, inferior inflow congestion, congested gastritis and a congested liver ascites and swelling

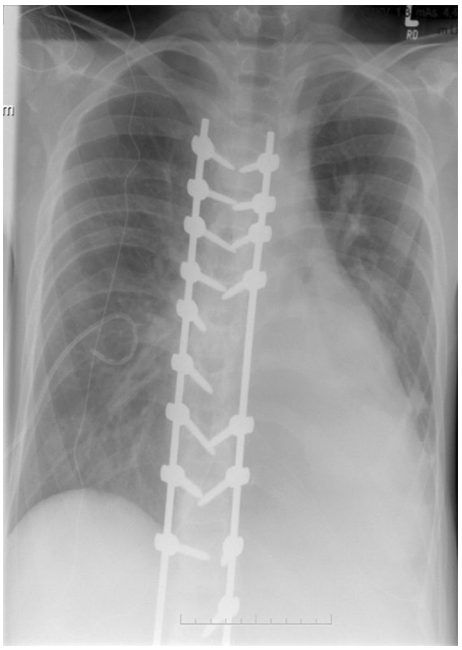

Fig. 5 X-ray showing the postoperative position after posterior spinal fusion. Cardiomegaly, uncertain pulmonary edema

of the lower limbs. Subsequently, the patient was transferred to the pediatric intensive care unit.

Therefore, the Nuss procedure was elected to solve the problems by achieving a sternal elevation by a sub-sternal bar used as an internal brace [6] to increase venous flow (Figs. 6, 7, 8, 9, 10).

The Nuss procedure was performed and a postoperative transthoracic echocardiography after showed an increased filling of the right ventricle and a regular flow of the inferior vena cava. During the clinical course, the volume status improved due to removed compression of the inferior vena cava with distinctive inferior inflow congestion. No complications occurred following the second surgical procedure, and the initial symptoms improved significantly during the clinical course. 3 weeks after the Nuss surgery, the patient was discharged with minimal residual pleural effusions and an oral diuretic therapy.

Discussion

Marfan syndrome is a congenital disorder of the connective tissue. It is an autosomal-dominant defect in microfibrils caused by mutations in the fibrillin-1 gene. This defect results in a weakness of elastic fibers. Marfan syndrome is a rare disease that affects both genders, with a prevalence of 1.5–17.2 per 100,000. Marfan syndrome is characterized by a wide variability in phenotypical appearances. In most patients, the cardiovascular, skeletal and ocular systems are

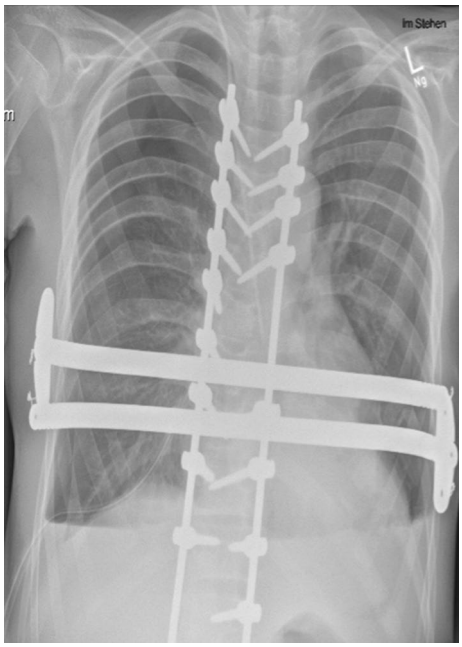

Fig. 6 X-ray showing the postoperative position after the Nuss procedure. Pleural effusions both sides. Thoracic drain right side

Fig. 7 Thoracic deformity prior to the Nuss procedure

affected by the disease. The diagnosis is on the basis of the wide variability of symptoms very complex [7, 8].

Inferior vena cava compression syndrome describes the clinical picture of reduced venous flow to the heart, which is also known as inferior inflow congestion. Pathophysiologic reasons can be the venous system itself, for instance, thrombosis or compression of the lower vena

Fig. 8 Thoracic deformity prior to the Nuss procedure

Fig. 9 Thorax after the Nuss procedure

Fig. 10 Thorax after the Nuss procedure

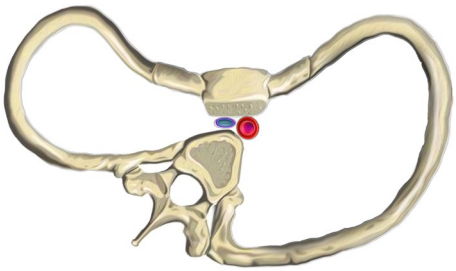

Fig. 11 Schematic illustration preoperatively

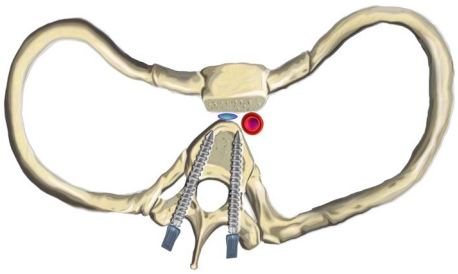

Fig. 12 Schematic illustration after posterior spinal fusion

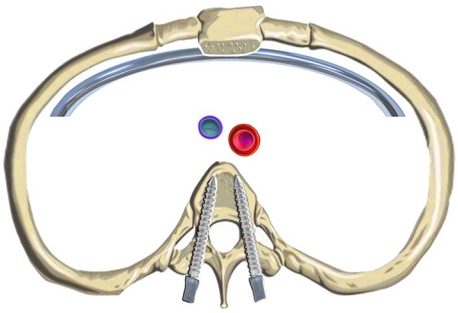

Fig. 13 Schematic illustration after Nuss procedure

cava by surrounding structures [9–11]. A clinical pertinent inferior vena cava compression syndrome, due to combined scoliosis and an extreme funnel chest, has not yet been described in the literature. Furthermore, no reference to a clinically pertinent inferior vena cava compression syndrome, among the wide range of cardiovascular symptoms in Marfan syndrome, can be found in the specialist literature. Ghazal et al. [12] described for the first time a congenital stenosis of the inferior vena cava in Marfan syndrome in their 2015 case report. Overall, congenital stenosis or aberrations of the inferior vena cava are very rare. Most commonly, stenosis of the inferior vena cava is observed in the area of the diaphragm or the liver [13].

In the present case, the preoperative cross-sectional imaging showed a significant constriction of the inferior vena cava close to the caval opening of the diaphragm (Figs. 2, 3); however, this was clinically asymptomatic. The correction of scoliosis resulted in a clinically relevant compression of the inferior vena cava against the distinctive funnel chest. Intraoperatively, a relevant cardiovascular depression was caused by the vena cava compression that was stabilized by mitigation of the reposition maneuver and intraoperative mass transfusion. During the clinical course, however, the symptoms of the inferior inflow congestion persisted, resulting in the urgent indication of a surgical correction of the distinctive funnel chest.

A treatment algorithm for the surgical procedure in distinctive scoliosis in the context of Marfan syndrome has not yet been described in the literature. The presented case shows that stenosis of the inferior vena cava already existed preoperatively; however, the clinical relevance concerning scoliosis correction is not yet apparent. The schematic illustrations show the compression of the inferior vena cava before and after each surgery (Figs. 11, 12, 13). MRI or multi-detector CT scans are used to detect this type of stenosis, although there are no data in the literature regarding the degree of restriction at which a clinically relevant inferior cava syndrome is expected. This is mainly due to the fact that the problem of congenital stenosis of the inferior vena cava in Marfan syndrome has not yet been sufficiently investigated.

Conclusion

When surgical scoliosis correction is indicated and planned in a patient with Marfan syndrome, scoliosis and funnel chest MRI or thoracic/abdominal CT should be performed preoperatively. When there is stenosis of the inferior vena cava, a multidisciplinary preoperative discussion including pediatric cardiologists, cardiothoracic surgeons, radiologists and orthopedic surgeons is recommended to evaluate the relevance of the stenosis. When a risk of further constriction of the inferior vena cava by surgery exists, it should be discussed whether a correction of the funnel chest should be performed first to prevent a clinically relevant compression syndrome. A standardized measurement of the diameter of the inferior vena cava should be carried out in all Marfan patients in order to detect individual risk.

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

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