

Case Report: Symptomatic Inferior Vena Cava Stenosis after Scoliosis Surgery in a Marfan Syndrome Patient with Pectus Excavatum

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Objectives: Marfan syndrome (MFS) is associated with a variety of cardiovascular symptoms, and deformities of the spine and the chest. We report a case of a 15-year-old boy with MFS, presenting with a symptomatic IVC compression after surgical scoliosis correction in the present of an asymmetric funnel chest (FC).

Methods/Results: A 15-year-old Caucasian MFS patient (aortic diameter $Z=4.54$, systemic score=9 and FBN1-positive) with a severe scoliosis and a FC underwent scoliosis surgery in an outside hospital. During the procedure the patient experienced severe hemodynamic instability, so that he was emergently transferred to our pediatric ICU. On arrival he was found to have severe ascites and extended edema of the lower limbs, which were resistant to diuretics. Imaging findings on a postoperative CT scan suggested a compression of the IVC in proximity to the right ventricle, between the sternum and the spine. Echocardiography demonstrated focal narrowing of the IVC with turbulent flow and a reduced diastolic filling of the right ventricle caused by a severe IVC constriction. Being a well-known patient in our interdisciplinary Marfan center, it was known that a previous MRI had shown a constriction of the IVC on the height of the diaphragmatic passage, which had been clinically asymptomatic at all times - an information that was not available at the outside hospital. Conservative treatment failed to mitigate the disastrous hemodynamic status caused by the inferior inflow congestion. A surgical decompression of the IVC via correction of the FC with a Nuss procedure was performed, leading to a significant improvement of the clinical symptoms. The clinical course proceeded without complications, and the patient was discharged after 3 weeks.

Conclusions: To our knowledge, this is the first case of symptomatic IVC compression in an MFS-patient after surgical correction of scoliosis. In the case of simultaneously existing FC and scoliosis in MFS, the FC correction should be performed before the scoliosis correction to avoid a hemodynamic deterioration. The indication for surgical corrections in MFS patients should be discussed in an interdisciplinary Marfan center, so that the full spectrum of the condition is taken into account.