

Straight Back Syndrome



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KEYWORDS

- Straight back syndrome • Tracheal obstruction • Chest wall deformity

KEY POINTS

- Straight back syndrome may rarely cause tracheal obstruction.
- Operative repair requires removal or thinning of the manubrium to relieve tracheal compression.
- Rarely the brachiocephalic artery also may be a cause of tracheal compression and require rerouting.

Straight back syndrome (SBS) is a rare congenital chest wall deformity such that the normal dorsal curvature of the thoracic spine is absent and instead is straight. This anatomic variant then effectively narrows the space between the posterior aspect of the manubrium and the anterior thoracic spine. This condition is often accompanied by pectus excavatum. If the loss of thoracic inlet space is severe enough, tracheal compression may ensue and lead to dyspnea. An additional cause of tracheal compression can be the brachiocephalic artery as it crosses the trachea in a narrowed space. Most patients with SBS do not have tracheal compression and do not exhibit respiratory symptoms but with extreme anterior spine displacement airway obstruction can occur.

SBS is most commonly associated with cardiac abnormalities, such as mitral valve prolapse and “pseudo-heart disease.”^{1,2} Patients can present with atypical chest pain, dyspnea, and palpitations. Approximately two-thirds of patients are found to have mitral valve prolapse either clinically or by echocardiography. One report described a patient with exertional pulmonary hypertension due to elevated pulmonary venous pressure due to compression of the left atrium and pulmonary veins from the narrow anterior-posterior chest.³

SBS causing airway compression severe enough to require operative treatment is so rare

that in general there are only occasional case reports. We reported 4 patients in conjunction with our French colleagues several years ago.⁴ In a general way, there are several possibilities to enlarge the space behind the upper sternum, including removing or thinning the manubrium, distracting the manubrium, elevating a severe pectus excavatum, or performing osteotomies of the anterior upper thoracic spine.

Each case is unique and an operative plan must necessarily be individualized. The computed tomography (CT) scan is the most important diagnostic modality and must be studied to identify points of airway compression and for possibilities for treatment. Our general philosophy was that it is less morbid to move the sternum than to remove the anterior portions of the offending vertebral bodies. Operations may be staged to gauge the results of an operative intervention if the initial procedure is thought likely to correct most of the airway compression. If the brachiocephalic artery is going to be divided and moved, it is prudent to study the circle of Willis to ensure that it is intact to minimize the risk of a watershed ischemic event.

Although each case is unique, a typical a classic case is illustrated in **Figs. 1–5**. The patient was a 21-year-old man with a long history of wheezing and dyspnea thought to be due to asthma. He had a mild pectus excavatum. His forced expiratory volume in 1 second was reduced at 37% of predicted. He eventually had a diagnosis of SBS.

Disclosures: None.

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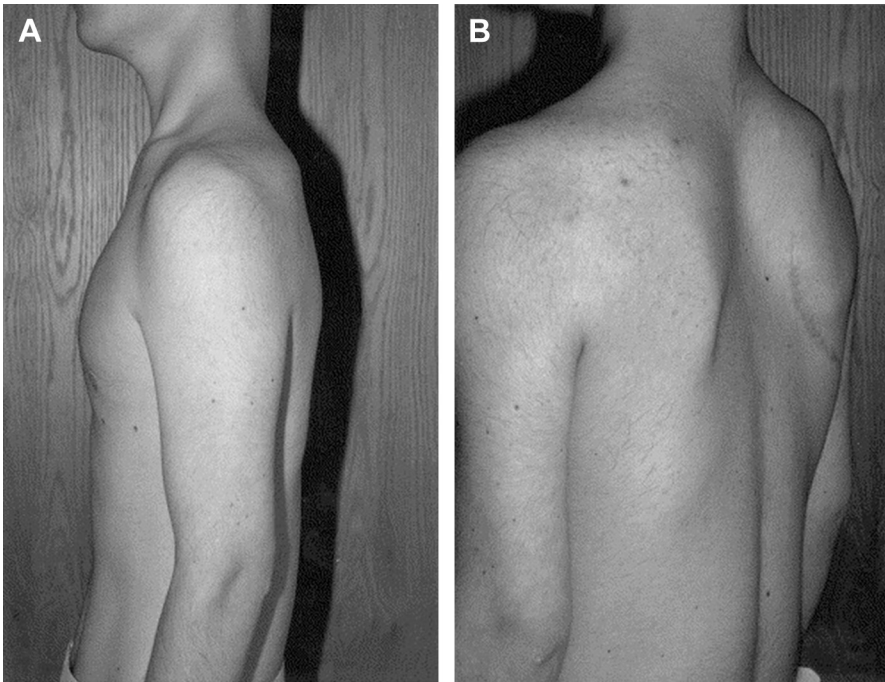


Fig. 1. A patient with SBS. Notice the narrow chest and the straight thoracic spine. (A) Lateral view (B). Posterior view. (From Grillo HC, Wright CD, Dartvelle PG, et al. Tracheal compression caused by straight back syndrome, chest wall deformity and anterior spine displacement: techniques for relief. *Ann Thorac Surg* 2005;80:2058; with permission.)

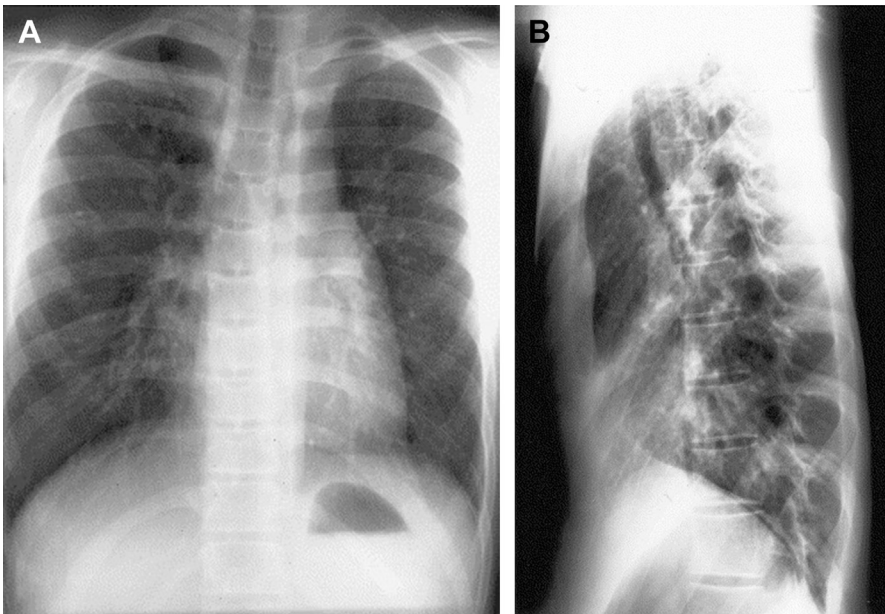


Fig. 2. A patient with SBS who required operation for tracheal compression (same as Fig. 1). (A) Chest radiograph anterior-posterior view demonstrating clear lung fields and slightly widened trachea. (B) Lateral view demonstrates a narrow chest with no dorsal curvature of the thoracic spine. (From Grillo HC, Wright CD, Dartvelle PG, et al. Tracheal compression caused by straight back syndrome, chest wall deformity and anterior spine displacement: techniques for relief. *Ann Thorac Surg* 2005;80:2059; with permission.)

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