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CLINICAL PRACTICE

## Scimitar syndrome: A rare explanation for () CrossMark a common symptom with an osteopathic approach



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## **KEYWORDS**

Scimitar syndrome; Partial anomalous pulmonary venous return; Dyspnea; Congenital heart defect; Osteopathic medicine; Osteopathic manipulation; Pneumonia

Abstract Background: Scimitar Syndrome is a rare congenital heart defect involving anomalous return of venous blood from the right lung. It is commonly associated with recurrent pulmonary infections, including pneumonia. Osteopathic manipulative treatment has been proven to improve the clinical course in hospitalized patients with pneumonia.

Clinical features: A 68-year-old female presented with dyspnea for 5 months. Examination disclosed decreased right-sided breath sounds and the presence of rightsided heart sounds. Osteopathic evaluation revealed both acute and chronic somatic dysfunctions and viscero-somatic changes. Chest computed tomography identified severe volume loss on the right with elevation of the right hemidiaphragm, a mediastinal shift towards the right and an anomalous venous drainage in the lower chest. Pulmonary angiogram confirmed Scimitar Syndrome. The patient developed pneumonia and was initiated on antibiotics. Osteopathic manipulation was implemented for symptomatic relief. The patient recovered quickly with a short hospital stay and symptom-free discharge.

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*Conclusions*: This case illustrates the importance of a thorough physical examination in combination with appropriate imaging in the discovery of a rare etiology for the treatment of a common complaint. Further, this is the first case describing the application of osteopathic manipulative medicine to aid in the diagnosis and treatment of the rare condition, Scimitar Syndrome. © 2015 Elsevier Ltd. All rights reserved.

Implications for clinical practice

- Despite being rare, partial anomalous pulmonary venous return should be considered in the presentation of a common pulmonary complaint.
- The use of osteopathic manipulation can more rapidly ameliorate symptoms and shortened hospital stay.
- Follow-up osteopathic manipulative treatment would likely enrich the patient's life by both reducing symptoms and extending time between recurrent pulmonary infections.
- Application of osteopathic manipulative medicine could be employed in the setting of rare anatomic anomalies, such as Scimitar Syndrome.
- Osteopathic evaluation may complement diagnosis and manipulative treatment may enhance physiologic function.

## Introduction

Partial anomalous pulmonary venous return (PAPVR) is an extremely rare family of congenital disorders constituting 0.5-1% of congenital heart disease.<sup>1</sup> Scimitar Syndrome, a subset of PAPVR, is even more rare, accounting for only 3-5% of all PAPVR cases<sup>2,3</sup> with an overall incidence of 0.002%.<sup>4</sup> Scimitar Syndrome involves the right lung being drained by a right pulmonary vein, denoted the scimitar vein, that is anomalously connected to the inferior vena cava (IVC).<sup>5</sup> This results in changes in the right lung, such that it becomes hypoplastic with atypical bronchial and vascular arrangement.<sup>5</sup> Draining of the anomalous pulmonary vein from the lung into the IVC produces a left-to-right shunt, which in-turn causes pulmonary hypertension and right ventricular overload; and thus commonly results in the development of right heart failure.

Symptom presentation depends upon the age of diagnosis, with the mean age of diagnosis being 7 months.<sup>6,7</sup> Infants have more severe symptoms and a poorer prognosis, tied directly to heart failure and pulmonary hypertension resulting in failure to thrive and cyanosis.<sup>8-10</sup> Approximately half of patients that are diagnosed after the infant stage remain asymptomatic, while others have symptoms ranging from fatigue and dyspnea to recurrent pneumonia. The predisposition for recurrent pulmonary infections is believed to lie with the changes in vascular anatomy, hypoplasticity of the affected lung, and changes in pulmonary pressure<sup>8,11</sup> Associated anomalies can include cardiovascular anomalies such as atrial or ventricular septal defect, dextrocardia, coarctation of the aorta, and patent ductus arteriosis along with pulmonary anomalies such as pulmonary sequestration, hypoplastic lung, and pulmonary vein stenosis. 1,4,6,9,12

Initial testing with chest radiography can show the classic anomalous vein shaped like a curved sword, providing the name "scimitar".<sup>2</sup> Doppler echocardiography demonstrates connections of the anomalous vein with the IVC or right atrium, as well as other associated changes to the heart.<sup>13</sup> Contrast computed tomography (CT) and magnetic resonance angiography (MRA) are the most useful in identifying and diagnosing the distinct anomalous vasculature.<sup>13</sup> Still, even with direct imaging, as this condition is so rare, it can often be missed.

Osteopathic manipulative medicine has been well established as having a role in the treatment of pneumonia.<sup>14</sup> The Multicenter Osteopathic Pneumonia Study in the Eldery (MOPSE) comprised a double-blinded, randomized, controlled trial evaluating the efficacy of osteopathic manipulative treatment as an adjunct in the treatment of pneumonia.<sup>15</sup> The MOPSE trial demonstrated that the use of osteopathic manipulative therapy significantly improved the outcome of pneumonia in the hospitalized patient by reducing length of stay, duration of intravenous antibiotics and associated respiratory failure or death.<sup>15,16</sup> With congenital structural changes resulting in Download English Version:

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