CASE REPORT

# Effective Long-Term Noninvasive Ventilation of a Woman With Kyphoscoliosis and Sinus Venosus Atrial Septal Defect

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**Atrial septal defects comprise one of the most common congenital cardiac malformations in adults and adolescents. However, such septal defects are rarely associated with chest wall diseases. When they are, respiratory failure soon develops and is severe. Cases in which the long-term course of disease is favorable with noninvasive ventilation have not been reported in the literature. We present the case of a 71-year-old woman with kyphoscoliosis, sinus venosus atrial septal defect, severe pulmonary hypertension, respiratory failure, and heart failure. We describe her response to treatment with noninvasive mechanical ventilation.**

**Key words:** *Kyphoscoliosis. Atrial septal defect. Noninvasive ventilation. Pulmonary hypertension.*

## Eficacia de la ventilación no invasiva a largo plazo en una paciente con cifoscoliosis asociada a comunicación interauricular tipo seno venoso

**Los defectos septales e interauriculares constituyen una de las anomalías congénitas cardíacas más frecuentes en adultos y adolescentes. Su asociación a enfermedades de la caja torácica es un hecho excepcional. Cuando ello ocurre, la aparición de insuficiencia respiratoria es precoz y de mayor gravedad. No hay casos descritos de evolución favorable a largo plazo con ventilación no invasiva. Presentamos el caso de una paciente de 71 años afectada de cifoscoliosis y de comunicación interauricular tipo seno venoso, con hipertensión pulmonar grave, insuficiencia respiratoria global e insuficiencia cardíaca, así como su respuesta al tratamiento con ventilación mecánica no invasiva.**

**Palabras clave:** *Cifoscoliosis. Comunicación interauricular. Ventilación no invasiva. Hipertensión pulmonar.*

## Introduction

Noninvasive mechanical ventilation (NIV), generally nocturnal, is used to treat chronic respiratory insufficiency due to restrictive disorders. Patients with sequelae of tuberculosis, kyphoscoliosis, hypoventilation-obesity syndrome, and neuromuscular disease are ideal candidates for receiving long-term NIV.[1] Respiratory insufficiency due to restrictive pulmonary disease can be exacerbated by other cardiorespiratory problems, such as atrial septal defects (ASDs), which constitute one of the most common congenital malformations in adults and adolescents.[2] ASDs have been associated with other congenital defects, and cases have been reported of autosomal dominant transmission associated with other malformations.[3] We report the case of a woman with kyphoscoliosis, an ASD of the sinus venosus type, severe pulmonary hypertension, global respiratory insufficiency, and heart failure—and describe her response to NIV. This is an exceptional case; the literature includes no reports of long-term favorable evolution with NIV.

## Case Description

The patient was a 71-year-old woman, allergic to beta-lactamase antibiotics and a noninsulin-dependent diabetic, who was diagnosed with moderate to severe kyphoscoliosis (Cobb angle, 48º). At 56 years of age echocardiography had led to a diagnosis of sinus venosus ASD and severe pulmonary hypertension. Until then she had been asymptomatic and had led a normal life. She was the mother of 2 healthy children. Surgical treatment of the defect was ruled out due to impaired lung function. At 62 years of age she started home oxygen therapy with nasal prongs for global respiratory insufficiency and mean pulmonary artery pressure greater than 50 mm Hg. While she was hospitalized due to progressive clinical

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Manuscript received August 11, 2005. Accepted for publication August 30, 2005.

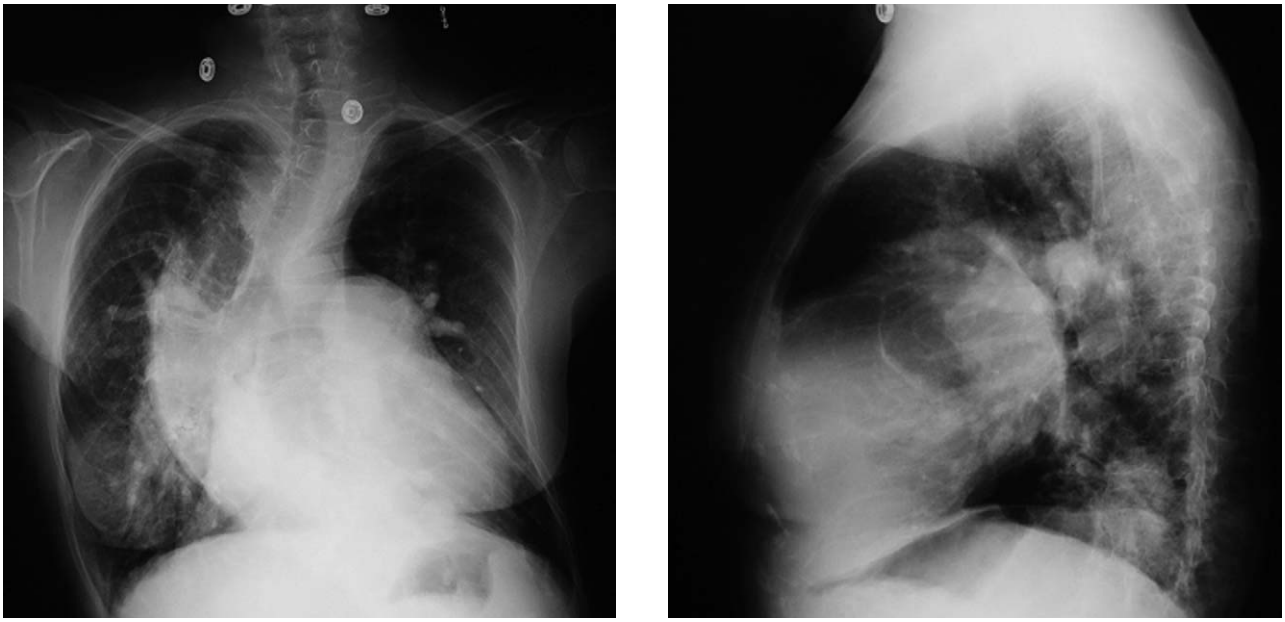

**Figure 1. Posteroanterior and lateral chest radiographs.**

deterioration and lack of response to oxygen therapy, she was classified as New York Heart Association (NYHA) functional class III. Physical examination revealed kyphoscoliosis with marked convexity to the right; cardiac auscultation revealed a 3/6 mesosystolic murmur with maximal intensity over the pulmonary and tricuspid area and an increased pulmonary component at the second sound. Lung function tests indicated a severe restrictive ventilatory defect: forced vital capacity was 960 mL (47%); forced expiratory volume in the first second, 778 mL (47%); and a ratio between those 2 values of 82%. Night pulse oximetry showed severe desaturation, with arterial hemoglobin saturation below 70% during the entire recorded period. Arterial blood gas analysis showed pH to be 7.37; $PaO_2$, 52 mm Hg; and $PaCO_2$, 87 mm Hg. Chest radiographs revealed marked kyphoscoliosis and cardiomegaly, mainly involving the right chambers and the left atrium, and prominence of the pulmonary cone with enlargement of the central and lobar arteries related to precapillary pulmonary hypertension (Figure 1). Transthoracic echocardiography revealed severe pulmonary hypertension (100/35 mm Hg), left-to-right shunt (Qp:Qs, 2.55), minimal systolic pericardial effusion on the posterior face of the left ventricle, a dilated right ventricle that showed thickened walls and severe disfunction and reversed atrial septal curvature, a defect of 14 mm in the atrial septum adjacent to the outlet of the superior vena cava indicative of sinus venosus ASD (Figure 2). NIV treatment was started, with a volumetric respirator and oxygen added to the circuit. At 3 days respiratory insufficiency had been corrected. For 8 years the patient has continued to use night ventilation with oxygen added to the respirator and has kept $PaO_2$ between 64 and 70 mm Hg and $PaCO_2$ within a normal range. The respirator settings for tidal volume ranged from 800 to 1000 mL; breathing frequency, at 20 breaths/min; and the inspiration:expiration ratio, at 1:1.5. Night pulse oximetry has shown arterial hemoglobin saturation to be higher than 92% during most of the recorded periods while the patient slept with the respirator; echocardiography showed high values for pulmonary pressure, with no change. During this 8-year period, the patient has required hospitalization on 3 occasions due to respiratory infection. She feels clinically stable (NYHA functional class II).

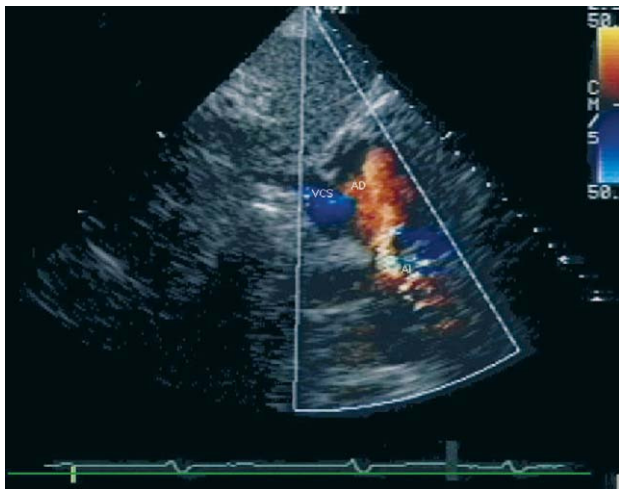


**Figure 2. Transthoracic color Doppler echocardiogram, subcostal projection, showing the outlets of both venae cava and the atrial septal defect. AD indicates right atrium; AI, left atrium; VCS, vena cava superior.**

## Discussion

ASD is commonly caused by an ASD in the oval fossa. In fewer than 10% of cases the defect is located posteriorly, outside the confines of the oval fossa, and is referred to as sinus venosus ASD. Reports of kyphoscoliosis associated with ASD are exceptional, and no cases are reported with long-term NIV treatment as in the present case. Defects of the sinus venosus type are usually silent and are therefore normally discovered

during adulthood. Transesophageal echocardiography is the only imaging modality that detects such defects and rules out coexisting partial pulmonary venous drainage abnormalities. It can be useful to inject a venous contrast medium to visualize the shunt.[4]

Henry et al[5] reported the case of a 73-year-old woman with ASD and kyphoscoliosis who was treated by percutaneous closure of the septal defect, which corrected the hypoxemia. In the present case, we ruled out surgical closure given the patient's high pulmonary pressure and severely altered lung function. Nevertheless, medical treatment and NIV with oxygen therapy enabled us to correct her respiratory insufficiency, improve her NYHA functional class from III to II, and achieve long-term stabilized pulmonary hypertension.

Survival of patients with ASDs is often relatively short although it is not uncommon for such patients to reach old age.[7] Mortality has been estimated to increase 6% annually starting at 40 years of age. One of the most severe complications is pulmonary hypertension, which appears in 15% of ASD patients and can progress rapidly. In such cases life expectancy is very short since the patients develop respiratory and heart failure with disease progression. In fact, our patient was developing respiratory insufficiency, but the course of disease was effectively controlled by NIV.[8] For 8 years the patient's arterial blood gas values have remained stable at their baseline values: $PaCO_2$ has remained within normal parameters; $PaO_2$, above 60 mm Hg. Likewise, there have been no changes in pulmonary hypertension.

Other less common complications in patients with ASDs are the development of atrial arrhythmia and, even less commonly, cerebral vascular phenomena and paradoxical embolisms—developments that did not occur in our patient.

The association of an ASD of the sinus venosus type with severe kyphoscoliosis is fairly rare. Both diseases contribute to the development of severe pulmonary hypertension. Such a scenario points in the direction of a poor short-term prognosis, even if closure of the defect is possible. An alternative to surgical or percutaneous closure for cases where such an option is ruled out could be optimization of medical treatment and the application of NIV.[1] Although our patient´s respiratory status was very serious, such a therapeutic approach led to an improvement in her functional class, correction of her respiratory insufficiency, and an increased life expectancy.

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