

# Sleep disordered breathing in kyphoscoliosis

JM Joshi\* and KD Modi\*\*

Department of Pulmonary Medicine,  
B.Y.L.Nair Hospital and T. N. Medical College, Mumbai 400008.

\* Professor \*\* Resident

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## Abstract

**Background:** Severe kyphoscoliosis may result in significant ventilatory defect, cardiorespiratory failure and death. Recent studies have suggested that sleep disordered breathing (SDB) may also contribute. Guilleminault and colleagues used the term "Quasimodo" syndrome for obstructive SDB in kyphoscoliosis after the famous hunchback of Notre Dame described by Victor Hugo. Contrary to these findings Sawicka and Branthwaite reported no evidence of obstructive SDB but only hypoventilation and oxygen desaturation during sleep in 21 subjects with non-paralytic and paralytic kyphoscoliosis. The authors concluded that higher body weight (57-71 Kg) and presence of systemic hypertension could have contributed to obstructive SDB in the Guilleminault study.

**Methods:** We analyzed our data of 11 cases of idiopathic kyphoscoliosis evaluated for SDB. The cases were referred in view of complaints of exertional dyspnoea during pre operative assessment for corrective spine surgery. A detailed history included respiratory symptoms, snoring and excessive daytime sleepiness (EDS). All patients were subjected to chest and thoracic radiograph, spirometry with flow volume loop (FVL), arterial blood gas analysis (ABG) and 2-dimensional echocardiography (2-D ECHO). Cobb's angle was calculated to assess the severity of scoliosis. Level 3 sleep study in the form of night- time recording of cardiorespiratory variables. Arterial CO<sub>2</sub> monitoring or end tidal CO<sub>2</sub> could not be performed. Titration study was performed with continuous positive airway pressure (CPAP) and bi-level positive airway pressure (PAP) on 2 separate nights. **Results:** Eleven patients, age range 13-50 years, 4 men and 7 women, bodyweight 21-43 Kg with kyphoscoliosis were studied. All had exertional dyspnoea, 3 complained of EDS while none snored. Four had previous history suggestive of right heart failure i.e. episodes of puffiness of face and pedal oedema. Cobb's angle was greater than 60 (70-110) degrees in all cases. All 11 cases had severe restrictive abnormality on spirometry and awake compensated type 2 respiratory failure i.e. hypoxaemia, hypercapnoea with a normal pH on arterial blood gas analysis. Four cases with symptoms of right heart failure were confirmed to have pulmonary hypertension and cor pulmonale on 2-D ECHO. Level 3 sleep study using cardiorespiratory variables showed nocturnal desaturations in one case while remaining 10 cases showed SDB in the form of hypoventilation and oxygen desaturations (hypoventilation/hypoxemia syndrome associated with sleep). While CPAP titration showed no improvement, titration study performed with bi-level positive airway pressure (PAP) showed improvement in hypoventilation and/or oxygen saturation. All cases were treated with bi-level therapy and showed improvement in symptoms and daytime arterial blood gases.

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*Address for correspondence*

**Dr. J. M. Joshi**, Professor and Head  
Department of Respiratory Medicine  
T. N. Medical College and  
B. Y. L. Nair Hospital, Mumbai-400008  
Tel: 022 23027642/43 Fax: 022 23003095  
Email: drjosijm@email.com drjosijm@gmail.com

## Introduction

Kyphoscoliosis is excessive lateral and anteroposterior curvature of the thoracic spine. [1] Up to 80% of cases with kyphoscoliosis are idiopathic with a female predominance of 4:1. [2] The severity of kyphoscoliosis is best determined by measuring Cobb's angle [3] which is defined as the angle between the perpendiculars of the lines drawn parallel to the upper border of the highest and lower border of the lowest vertebrae. Severe kyphoscoliosis may lead to a significant ventilatory defect, cardiorespiratory failure and premature death. [4] Although, some studies have shown lung volume abnormalities with spinal curvatures of 30° to 40° [5] probably related to poor respiratory muscle strength [6,7], Weinstein et al [8] showed that Cobb's angle could be correlated with severity of lung function abnormality and a Cobb's angle above 60° is associated with progressive cardiorespiratory failure. Mirhead et al [9] have also shown that restrictive disorder and hypoxia in kyphoscoliosis are linked to the severity of the deformity. Factors contributing to respiratory failure are thought to include reduced surface area for diffusion, ventilation-perfusion inequality, alveolar hypoventilation, increased respiratory work, respiratory muscle weakness and an abnormal increase in pulmonary artery pressure during exercise. [10,11]

Ventilation is already impaired in the awake patient with kyphoscoliosis. Further impairment of ventilatory mechanics in sleep may cause significant sleep disordered breathing (SDB). [12] Guilleminault et al [13] in a series of 5 cases showed a complex SDB associated with obstructive apnea and hypopnoea. They coined the term Quasimodo syndrome for SDB associated with kyphoscoliosis after the legendary hunchback of Notre Dame described by Victor Hugo. Contrary to these findings in a subsequent study of 11 subjects with non-paralytic and 10 with paralytic kyphoscoliosis; Sawicka and Branthwaite reported only hypoventilation and oxygen desaturation during sleep but no obstructive SDB. [14] We therefore present data of 11 cases of kyphoscoliosis with SDB seen in our department.

## Patients and Methods

We retrospectively analysed 11 cases of idiopathic kyphoscoliosis with SDB. These cases were referred for preoperative assessment prior to corrective spine surgery in view of complaints of exertional dyspnoea. A detailed

history included respiratory symptoms, snoring and excessive daytime sleepiness (EDS). All patients were subjected to chest and thoracic radiograph, spirometry with flow volume loop (FVL), arterial blood gas analysis (ABG) and 2-Dimensional echocardiography (2-D ECHO). Cobb's angle was calculated to assess the severity of scoliosis. Level 3 sleep study in the form of night-time recording of cardiorespiratory variables which included airflow with oro-nasal flow sensors, snoring by microphone, oxygen saturation and heart rate by pulse oximetry, thoraco-abdominal movements and body position by sensors. Arterial CO<sub>2</sub> monitoring or end tidal CO<sub>2</sub> were not performed. Titration studies were done with continuous positive airway pressure (CPAP) and bi-level positive airway pressure (bi-level PAP) on separate nights.

## Results

Eleven patients, age range 13 - 50 years, 4 men and 7 women, bodyweight 21-43 Kg with kyphoscoliosis and exertional dyspnoea were studied. Three complained of EDS while none snored. Four had previous history suggestive of right heart failure i.e. episodes of puffiness of face and pedal oedema. Cobb's angle was greater than 60 degrees in all cases-70-110 degrees. Spirometry showed severe restrictive abnormality in all and none showed upper airway obstruction on the FVLs. ABG showed compensated type 2 respiratory failure i.e. hypoxaemia, hypercapnoea with a normal pH in all cases. Four cases with symptoms suggestive of right heart failure showed evidence of pulmonary hypertension and cor pulmonale on 2-D ECHO. On limited sleep study, 10 cases had hypopnoeas with oxygen desaturations (figure 1) -indicative of hypoventilation [14]; while one had only oxygen desaturations. Thus all cases showed hypoventilation/ hypoxemia syndrome associated with sleep [15]. Although hypoventilation is defined as an increase of PaCO<sub>2</sub> levels  $\geq 10$  mm Hg during sleep compared to PaCO<sub>2</sub> levels in wakefulness or in supine position, sleep studies do not routinely include arterial PaCO<sub>2</sub> as it is invasive and the surrogates for PaCO<sub>2</sub>, such as end-tidal PCO<sub>2</sub> monitoring and transcutaneous capnography are inaccurate [15]. Therefore in the present study even though PaCO<sub>2</sub> monitoring was not performed; hypoventilation was diagnosed based on successful correction of nocturnal O<sub>2</sub> desaturation and chronic hypercapnoea by nocturnal ventilation using bi level PAP (and not CPAP), in accord with the American

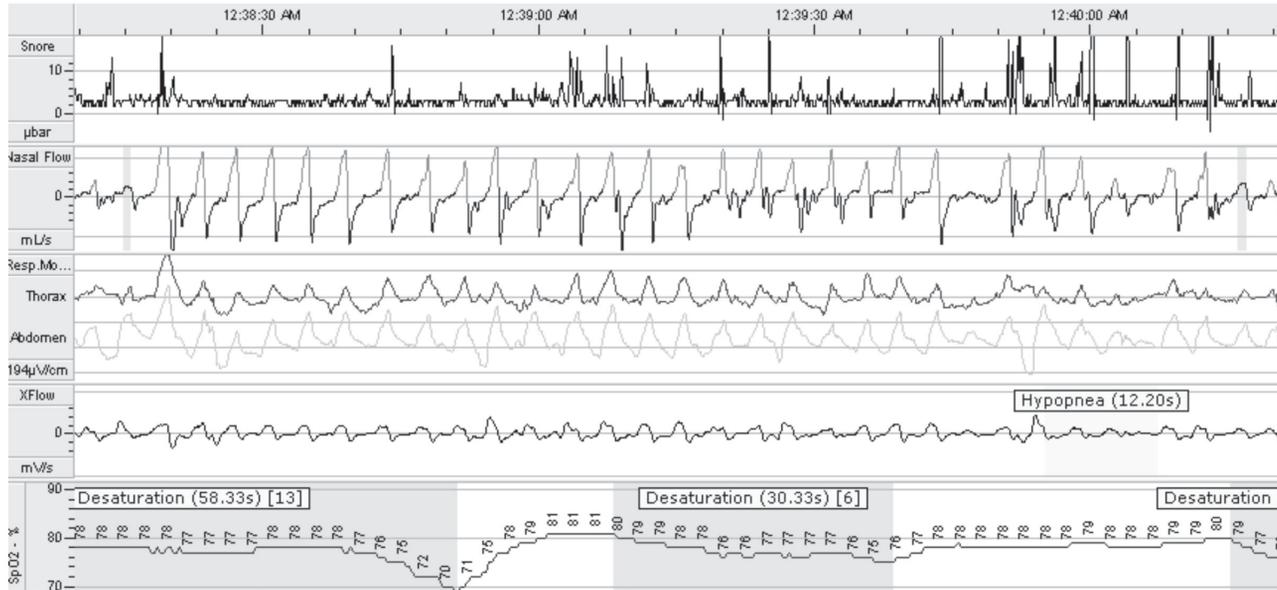


Figure 1: Level 3 limited Sleep study using cardiorespiratory channels showing hypopnoea with oxygen desaturations.

Academy of Sleep Medicine (AASM) recommendations [16].

**Discussion**

There is very little published data on SDB in kyphoscoliosis [12,13,14]. This, along with data of the present series is summarized in table 1. Mezon et al [12] first reported a spectrum of sleep related breathing abnormalities in five patients with severe kyphoscoliosis. The abnormalities ranged from no disorder to severe episodes of prolonged central apnea. Rapid-eye-movement (REM) sleep was the period of greatest physiologic disturbance in all the patients, being the time for greatest oxygen desaturations. Guilleminault et al [13] reported obstructive SDB in five kyphoscoliotic subjects

all of whom were relatively overweight, 3 had overt symptoms of sleep apnoea and several had associated systemic hypertension. While all our cases were low-normal weight, none snored, only three had EDS and none had systemic HT. All were symptomatic with exertional dyspnoea, Cobb's angle of greater than 60 degrees, severe restrictive abnormality, hypoxaemia and hypercapnoea on ABG. Ten out of our eleven patients had nocturnal hypoventilation with oxygen desaturation consistent with the findings of Mezon et al [12] and Sawicka & Branthwaite [14]. Unlike the cases of Guilleminault series there were no obstructive SDB. However, obstructive SDB in the Guilleminault series can be explained by their heavier body weight (57-71Kg) and presence of systemic hypertension, both conditions known to predispose to obstructive events. While Mezon

**Table 1: Sleep Disordered Breathing in Kyphoscoliosis: Comparative Data of Previous Published Studies and the Present Study**

Study [Ref]	No	Age	Weight	Kyphoscoliosis (Severity)	Complications (Awake)	Type of SDB
Mezon et al [7]	5	NA	NA	Severe	Hypoxaemia, polycythemia, Pulmonary HT, Cor Pulmonale	None, central sleep apnoeas
Guilleminault et al [8]	5	43-62	57-71	90°-115°	Systemic HT	Obstructive apneas/hypopnoeas
Sawicka & Branthwaite [9]	21	11-57	16.3-60	60°-140°	Hypoxaemia, increase end tidal CO2	Hypoventilation, O2 desaturation
Present Study	11	13-50	21-43	70°-110°	Hypoxaemia, hypercapnoea, Pulmonary HT, Cor Pulmonale	Hypoventilation, O2 desaturation

et al found no apparent relation of derangements in breathing pattern to the degree of thoracic deformity, pulmonary function tests or ABG, Sawicka & Branthwaite reported nocturnal hypoventilation in their series of paralytic and non paralytic kyphoscoliosis all having awake hypoxaemia and increased end tidal CO<sub>2</sub>. All 11 cases in our series with hypoventilation/ hypoxemia syndrome associated with sleep were low to normal body weight (21-43 Kg), none had systemic hypertension but all had awake blood gas abnormality similar to cases in the Sawicka & Branthwaite series. Therefore, weight, systemic hypertension and awake blood gas abnormalities seem to be a predictor of presence and type of breathing abnormality during sleep in kyphoscoliosis.

Four of our patients with SDB had features suggestive of cor pulmonale. Sleep-induced respiratory abnormalities with concomitant hypoxemia, hypercarbia, and acidosis may participate in the deterioration of the cardiovascular status of patients with kyphoscoliosis, leading ultimately to cor pulmonale and right ventricular failure. [13] Cases with clinical evidence of chronic hypoxemia, polycythemia, and cor pulmonale are thus likely to have most severe derangements during sleep as previously reported by Mezon et al. This occurs due to the increased elastic load of the stiff chest wall and diminished neural drive to the intercostals muscles during sleep. The burden of expanding the non distensible chest wall falls on the diaphragm, [17] The severity of sleep related breathing disorders in kyphoscoliosis thus can be determined by the magnitude of blood gas derangements and presence of cor pulmonale.

There was good response to treatment with bi-level PAP in all our cases, with reduction in symptoms, improvement in hypoventilation, oxygen desaturation and daytime hypercapnoea. Similar results have been reported by Gonzalez et al [18] who have shown that long-term NIPPV therapy improves daytime blood gas levels, respiratory muscle performance, and hypoventilation-based symptoms in patients with severe kyphoscoliosis. In another study [19] Buyse et al showed that nocturnal nasal intermittent positive pressure ventilation (NIPPV), plus long term oxygen therapy results in more favourable survival and changes in blood gases and respiratory function than long-term oxygen therapy alone.

When kyphoscoliosis is complicated by cardiorespiratory failure, the prognosis is grim. [20] However, sleep-related breathing disorders represent a potentially treatable cause

of respiratory failure. In addition to patients who are overweight and hypertensive, cases of severe kyphoscoliosis who have awake blood gas abnormalities must be evaluated for SDB. Bi-level positive airway pressure therapy is the treatment of choice in hypoventilation/ hypoxemia syndrome associated with sleep in kyphoscoliosis.

## Conclusions

SDB represent a potentially treatable cause of cardiorespiratory failure in kyphoscoliosis. In addition to patients who are overweight and hypertensive, cases of kyphoscoliosis who have awake arterial blood gas abnormality must be evaluated and treated for SDB. Bi-level positive airway pressure therapy is the treatment of choice.

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