Case Report

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# Kyphoscoliosis complicating asthma with fixed airway obstruction

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ABSTRACT

**Introduction:** Kyphoscoliosis is present in up to 2% of the juvenile population and can have deleterious effects on respiratory mechanics, leading to chronic respiratory failure later on in adult life.

**Case presentation:** Hereby we describe a 53-year-old patient with severe uncontrolled asthma who presented with chronic hypercapnic respiratory failure. During her medical workup, she was noted to have several comorbidities leading to her respiratory failure. The patient had radiological evidence of bronchiectasis with recurrent episodes of infection, and a severe deformity of the spine due to Kyphoscoliosis. Probably the kyphotic component of this deformity had worsened due to a long history of oral steroid use leading to severe osteoporosis and consequent vertebral compression fractures reaching a Cobb angle of 73 degrees. This was probably caused by the patient's non-compliance with inhaler therapy and an excessive reliance on oral steroid use. Her respiratory failure was treated with domiciliary noninvasive positive pressure ventilation and 24-hour oxygen therapy and her symptoms improved.

**Conclusion:** A multidisciplinary approach across different specialities is necessary when managing such a patient with kyphoscoliosis, bronchiectasis, asthma with airflow limitation with respiratory failure.

**Key words:** Kyphoscoliosis, Respiratory failure, Non-invasive ventilation

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## Introduction

Kyphoscoliosis is defined as "a deviation of the normal curvature of the spine in the sagittal and coronal planes and can include a rotation of the spinal axis"[1, 2] . Adult scoliosis is defined as a lateral deviation of more than 10 degrees in the lateral plane, and kyphosis in the sagittal plane as measured by the Cobb angle, which is formed by the intersection of two lines, one parallel to the top and the other parallel to the bottom vertebrae of the scoliotic or kyphotic curves [1, 3]. We present a 53 year old patient with severe uncontrolled asthma who presented with chronic hypercapnic respiratory failure.

## Case report

A 53-year-old woman presented with a 3-day history of worsening shortness of breath on walking a few metres on the flat associated with a cough productive of a large volume of greenish sputum and fever.

On examination, the patient's respiratory rate was 16 breaths/ minute, she had a pulse rate of 110 beats/ minute with a temperature of 39 degrees Celsius, with an oxygen saturation of 85% on air. Auscultation of the chest revealed diffuse wheezing in both lung areas. Arterial blood gases taken while the patient was on oxygen concentration of 28% via venturi mask showed pH 7.34, $PaCO_2$ 86 mmHg, $PaO_2$ 75.1 mmHg, oxygen saturation 93%, lactate 0.6 mmol/litre, and $HCO_3^-$ 48.2 mmol/µl.

On examining the patient's notes, it was noted that she had also suffered from asthma for the previous 20 years and she had been prescribed a fluticasone 250mcg inhaler 2 puffs twice daily and salmeterol 25 mcg inhaler 2 puffs twice daily via metered dose inhaler (pmdi). She had also been prescribed 2 puffs of 100mcg salbutamol on an as-required basis via pmdi. Total IgE was 80 international units/ml (Range 0-30), while IGE to House dust mite 1.58iu/ml (0-0.34) Cat 0.45(0.034). Eosinophil count 0.04(0-0.06x$10^{-9}$per l). An HRCT thorax taken two years before admission (Figure 1) had shown evidence of Bronchiectasis which had already been detected by another HRCT 6 years previous to the first. Regular treatment included prophylactic azithromycin 500mg three times a week.

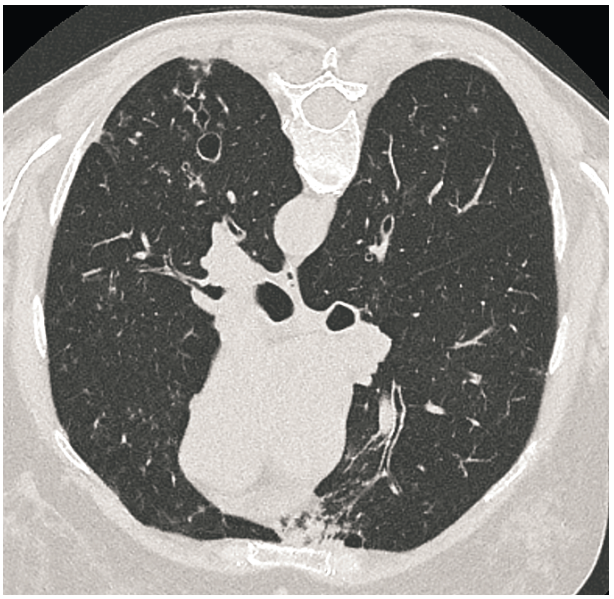


**Figure 1.** HRCT showing non tapering bronchioles with Signet ring appearance and classic tram track sign appearance of bronchi anteriorly, in keeping with bronchiectasis.

Notwithstanding, she had been persistently symptomatic with frequent visits to her family doctor, or the emergency room up to 3-4 times per month. General practitioner and hospital outpatient notes expressed the opinion that the patient had been poorly compliant with inhaler therapy and often resorted to Beta-agonists via inhalers or nebulizers. She also required regular courses of oral steroids 3-5- times a year. She had also refused a trial of treatment with anti-IgE therapy omalizumab, and was not considered for anti-IL5 therapy because of the low eosinophil counts.

Her lung function tests which included plethysmography and spirometry with a bronchodilator challenge at a medical outpatients (MOP) visit, 28 months before this presentation, were as per Table 1. The $FEV_1$

**Table 1.** Plethysmography results 2 years prior to admission.

| | | | % predicted |
|---|---|---|---|
| $FEV_1$ | Volume | 0.48 Litre | 21% |
| FVC | Volume | 1.28 Litre | 43% |
| $FEV_1$/FVC | Ratio | 0.38 | |
| TLC | Volume | 6.45 Litre | 143% |
| Residual Volume | Volume | 5.16 Litre | 343% |
| RV/TLC | Ratio | 0.8 | |

and FVC were 21% and 43% of the predicted value respectively. $FEV_1$/FVC ratio of 38% suggesting a diagnosis of COPD, and labelled GOLD 4. On plethysmography which was performed on a separate occasion, the Residual Volume was significantly high both when compared to the predicted value and as a fraction of the total lung capacity. A reversibility of 17% of $FEV_1$ was noted on lung function performed on another follow up visit.

Routine blood tests on admission were unremarkable with a C reactive protein level of 40mg/L. A respiratory screen for common respiratory viruses and bacteria (including *Influenza A, B and C, corona NL63, 229E, OC43, HKU1, Metapneumo virus A* and *B, adenovirus, rhinovirus, respiratory syncytial A* and *B, Mycoplasma Pneumonia, Haemophilus influenzae B, Strep. Pneumoniae, Chlamydia Pneumoniae, Klebsiella Pneumoniae, Legionella Pneumophilia/Longbeachae, Moraxella Catarrhalis, Bordatella species, Staphyloccus aureus*, and COVID-19) was negative, as well as a specimen of sputum which also cultured no bacteria.

A chest x-ray (Figure 2) was reported as showing airspace shadowing possibly as result of right lower zone consolidation and the presence of kyphoscoliosis was suspected.

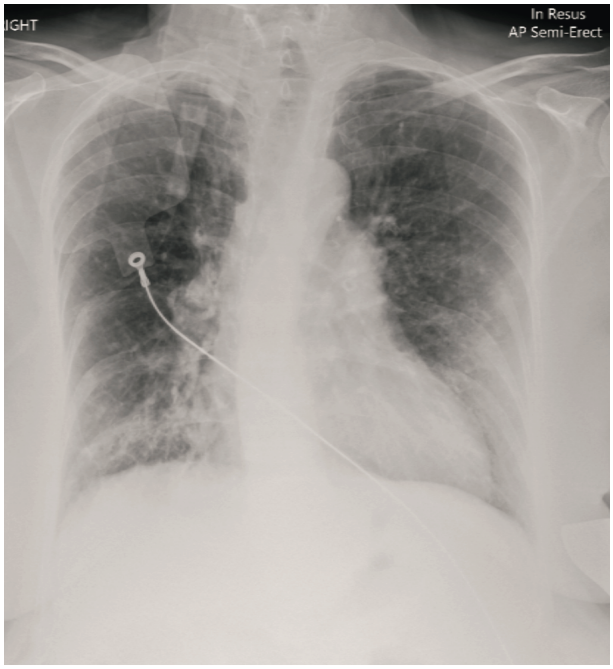


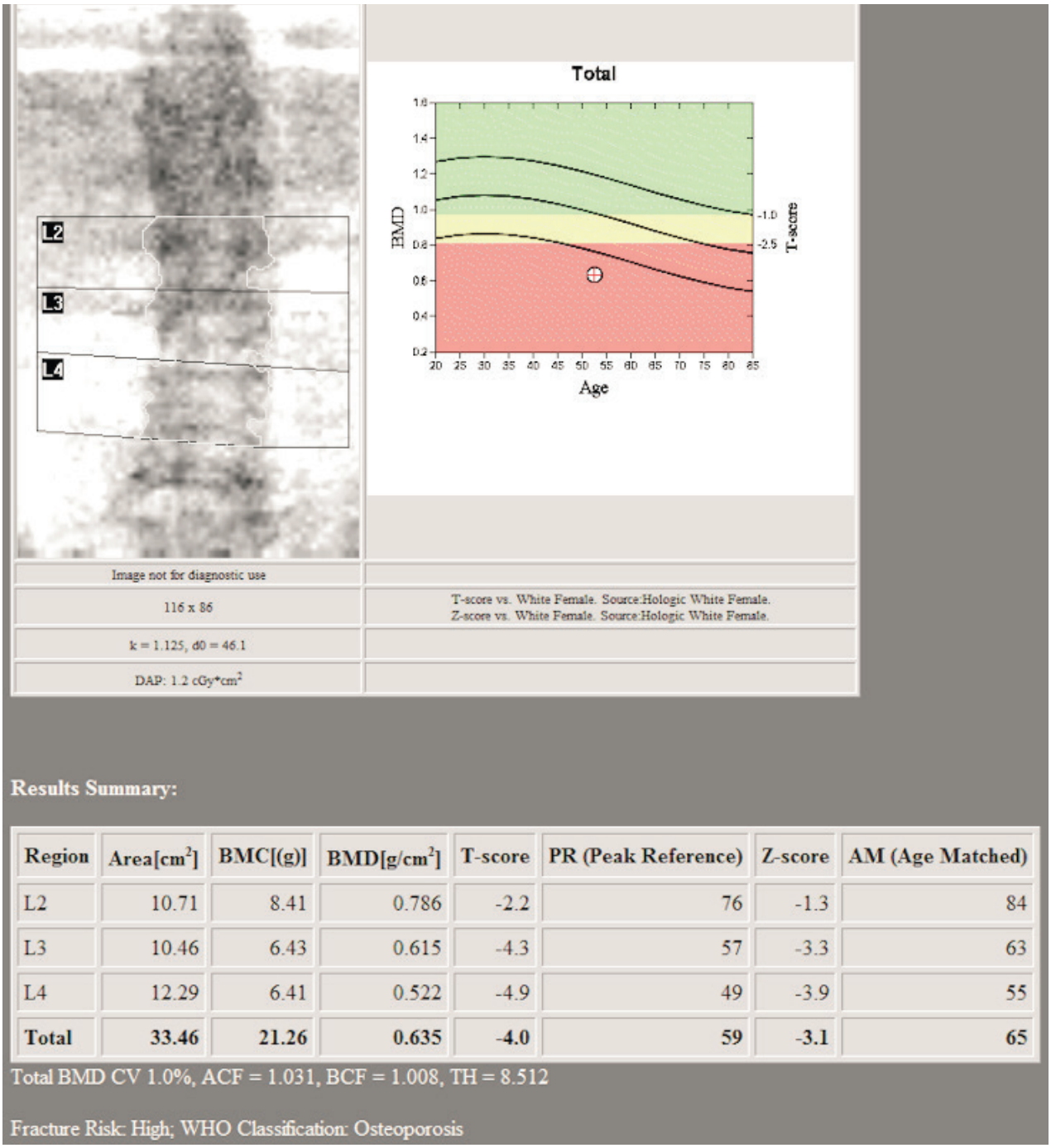

**Figure 2.** Chest X-ray taken on admission showing airspace shadowing in the right lower zone consolidation and the presence of kyphoscoliosis.

She was started on 28% oxygen and given 5mg of salbutamol and 0.5mg ipratropium bromide via nebulizer , followed by 100mg IV hydrocortisone, 4.5g of piperacillin/tazobactam IV three times daily and a single infusion of intravenous magnesium sulphate of 2g over 30 minutes. She was immediately started on non-invasive ventilation for her acute on chronic hypercapnoeic respiratory failure. She was admitted to hospital where her clinical condition, as well as her arterial blood gas results (Table 2), improved over the next few days; however she remained breathless on exertion on walking around 20m. At that point she was discharged from hospital on a tailing down dose of prednisone and her regular inhaler therapy. Bilevel domiciliary NIV was started at 16 cm IPAP and 5 cm EPAP during the night. She was also prescribed 24-hour oxygen therapy capped at 28% oxygen.

A rheumatology review was organized. A previous bone density result which had been performed a few months before admission showed evidence of severe osteoporosis with a T score of -4.0 and collapse of a number of thoracic vertebrae (Figure 3). This was considered to be most likely due to years of recurrent courses of oral steroids by the patient. In view of this, she was prescribed a yearly 15mg dose of intravenous zoledronic acid, a bisphosphonate. Kyphoscoliosis was confirmed on a scoliogram which reported a right convex thoracic component with a Cobb angle of approximately 19 degree and a left convex lumbar component with a Cobb angle of 16 degrees (Figures 4 and 5). Kyphosis was confirmed on lateral chest x-rays which showed a Cobb angles 73 degrees (Figure 6)..

**Table 2.** Arterial blood gas results prior to and after initiation of NIV therapy

| | **ABG during exacerbation on $O_2$ via 28% Venturi mask (prior to iniation of NIV therapy)** | **ABG on NIV** | **Reference Range** |
|---|---|---|---|
| pH | 7.34 | 7.46 | 7.35 - 7.45 |
| $pCO_2$ | 86 mmHg | 43.9 mmHg | 35 - 45 mmHg |
| $pO_2$ | 75.1 mmHg | 66.1 mmHg | 80 - 100 mmHg |
| $SO_2$ | 93% | 94.5% | 95 - 99% |
| $HCO_3^-$ | 48.2 mmol/L | 37.2 mmol/L | 22 - 26 mmol/L |




| Region | Area[cm²] | BMC[(g)] | BMD[g/cm²] | T-score | PR (Peak Reference) | Z-score | AM (Age Matched) |
|---|---|---|---|---|---|---|---|
| L2 | 10.71 | 8.41 | 0.786 | -2.2 | 76 | -1.3 | 84 |
| L3 | 10.46 | 6.43 | 0.615 | -4.3 | 57 | -3.3 | 63 |
| L4 | 12.29 | 6.41 | 0.522 | -4.9 | 49 | -3.9 | 55 |
| **Total** | **33.46** | **21.26** | **0.635** | **-4.0** | **59** | **-3.1** | **65** |

**Figure 3.** Bone densitometry result showing evidence of osteoporosis with T-score of -0.4.

Unfortunately, compliance with NIV therapy was also an issue and she relied mostly on oxygen therapy via concentrator.

## Discussion

On reviewing her case, multiple factors were considered to be contributing to her type 2 respiratory failure. First of all, the patient had been diagnosed

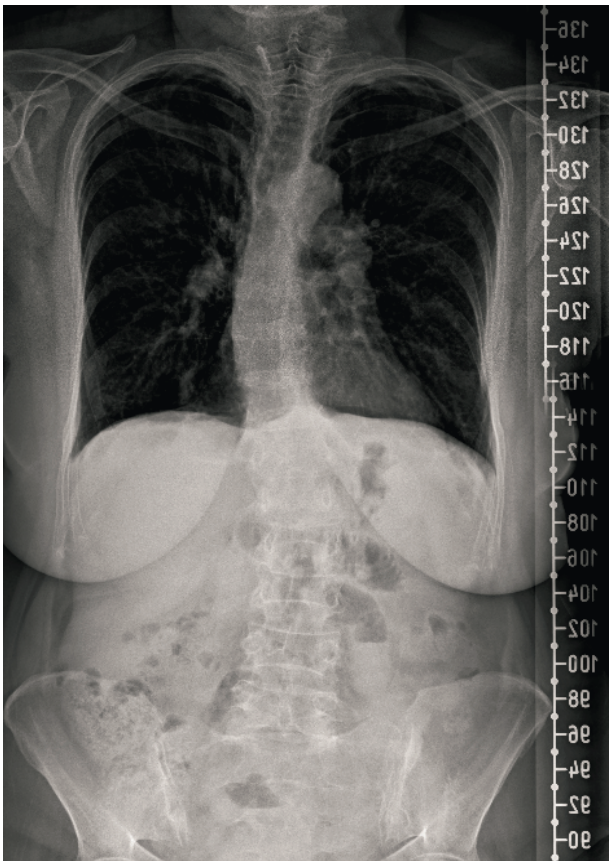


**Figure 4.** Scoliogram showing thoracolumbar sigmoid scoliosis, with a right convex thoracic component with a Cobb angle of approximately 19 degree and a left convex lumbar component with a Cobb angle of 16 degrees.

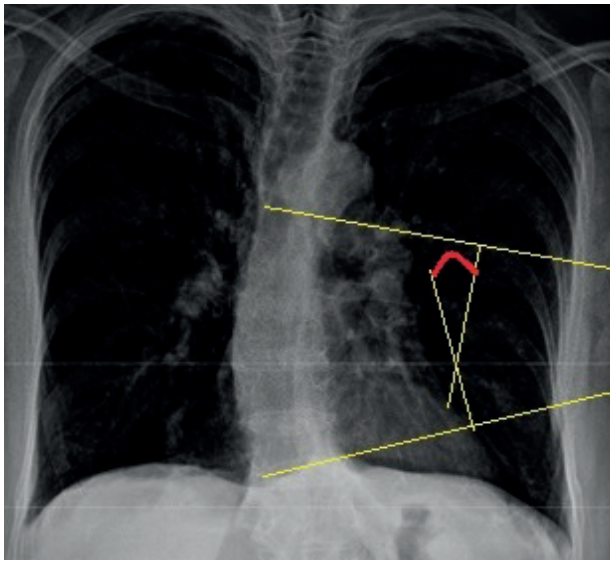


**Figure 5.** Chest X-ray showing thoracic scoliosis with a Cobb angle of 19 degress.

with asthma over 20 years before. Her medical notes revealed that previous caring physicians had expressed the opinion that the patient was poorly compliant to her regular preventer inhaler therapy and often resorted to beta agonist via inhalers or nebulizers. She also had a longstanding history of regular exacerbations requiring multiple courses of oral steroids and antibiotics.

The Pulmonary function tests suggested that the patient might have suffered from COPD Stage 4 [4]. As the patient was a non-smoker this obstructive airway deficit was more likely to be secondary to long-term uncontrolled asthma, which GINA guidelines more appropriately describe differently as airway remodeling and refer to it as "Asthma with airflow limitation" [5].

Asthma patients with fixed airway obstruction have a worse disease course with increase in symptoms, exacerbations and higher mortality. These patients exhibit a progressive decline in $FEV_1$ and an exacerbation rate similar to patients with COPD, which is indeed higher than that of asthma patients with no fixed airway obstruction [4, 5].

The presence of bronchiectasis was probably both the consequence of respiratory infections but also a frequent trigger for subsequent acute asthma attacks, which created a vicious cycle with a progressive loss of lung function with every episode as described by Ni Y et al [6]. The diagnosis satisfied the criteria for the definition by the European Respiratory Society guideline on Bronchiectasis as, "a chronic respiratory disease

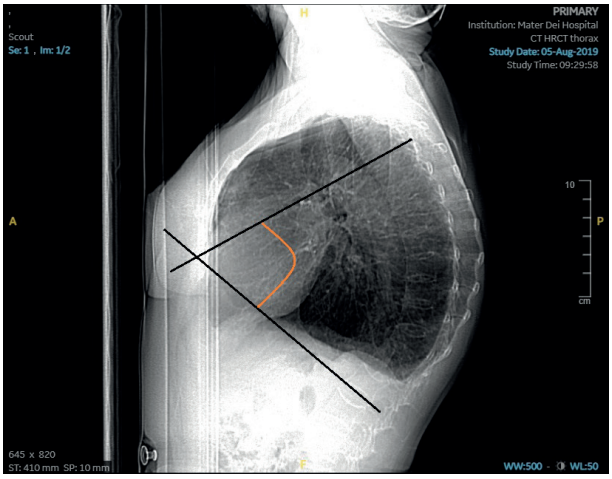



**Figure 6.** Lateral Chest x-ray showing thoracic kyphosis with a Cobb angle of 73 degrees.

characterized by a clinical syndrome of cough, sputum production and bronchial infection, and radiologically by abnormal and permanent dilatation of the bronchi" [7]. The HRCT taken both at 9 and 3 years prior to the admission both had clearly documented classical "Signet ring" and "tramline" radiological signs of bronchiectasis (Figure 1).

Furthermore, the patient had confounding restrictive lung disease caused by a severe kyphoscoliosis. The patient probably may have had initially a moderate form of idiopathic kyphoscoliosis which was further aggravated over many years by the development of severe osteoporosis, likely to be due to the frequent courses of oral steroids which greatly accentuated the kyphotic curves as indicated by the cobb angle of 73 degrees of the kyphotic curve (Normal <10 degrees). While NIV was successful in this patient during the acute episode, Adıgüzel et al. report that 20% of patients with kyphoscoliosis admitted to intensive care for acute respiratory failure needed intubation where accompanying sepsis was the main reason [8].

Most cases of kyphoscoliosis in adults are idiopathic with genetic, epigenetic, and environmental contributors. It is estimated that up to 2% of the adolescent population may have some degree of kyphoscoliosis [9], amongst whom only 0.4% result in significant clinical problems [9]. Idiopathic kyphoscoliosis has mostly a benign clinical course [1]. However, many cases of kyphoscoliosis are not idiopathic but are the result of loss of muscular support caused by neurological disease, for example muscular dystrophy, cerebral palsy or motor neuron disease [10]. This second possibility was clearly not the cause in this patient and will not be discussed in this paper.

Many countries have school screening programmes for children and adolescents aged 10-18 for the presence of kyphoscoliosis. The US Preventive Services Task Force (USPSTF) concluded that "current evidence is insufficient to assess the benefits and the harms of this screening" [11]. However, it determined that the concurrent of use of the forward bend test, the scoliometer and Moire topography had a 93.8% sensitivity and 99.2% specificity to detect idiopathic scoliosis" [11]. While bent over, palms pressed together and arms dangling, the scoliometer (inclinometer) measures the curvature of the patient's spine, providing objective measurement for possible referral [9]. While Moire imaging by projecting 3D images on the trunk is useful, radiographic examination of the spine remains the gold standard [12].

In adolescent idiopathic scoliosis bracing is recommended for patients with a Cobb angle between 25-45 degrees [9, 13]. There are two types of braces either soft or rigid including 25 different designs [13]. Surgery is reserved only for immature skeletons with a Cobb angle >45 degrees in adolescents; however the description of the various techniques is outside the scope of this paper [9]. The USPSTF found adequate evidence to justify the use of braces, and inadequate evidence for treatment with exercise and surgery [11].

Physical therapy has also been suggested to be helpful in adolescent kyphoscoliosis, in a systematic review of the literature by Gonzales-Galves et al., and suggested that strengthening was more important than stretching in kyphosis while both are important for lordosis [14]. They also conclude that randomized controlled studies are necessary to establish which are the best exercises [14].

Severe Kyphoscoliosis leads to diminished chest wall compliance and impaired respiratory mechanics, leading to progressive hypoventilation, chronic hypercapnic respiratory failure, and hypoxemia [8]. Lung function tests are of a restrictive pattern, with a decrease in total lung capacity and vital capacity, but normal residual volume and a resultant increase in the RV/TLC ratio [1, 2]. In the case reported here the residual volume was greatly increased probably because of severe obstructive airways disease, while the Forced Vital Capacity was greatly decreased by both diseases. In fact the RV/TLC ratio was greatly increased as shown on Table 1.

A functional assessment should be included in these patients. In fact, a 6-minute walk test has been shown to predict the degree of limitation in respiratory function better than pulmonary function tests or arterial blood gases [15]. This was not considered necessary in this case because the patient had dyspnoea on minimal exertion and abnormal arterial blood gases at rest.

Bergofsky et al. in his classical description of 1959 reported three main anatomical changes in the lungs of kyphoscoliosis patients, namely chronic pulmonary

emphysema with bronchial obstruction, changes in the small vessels of the lung, and tangled and compressed vessels. The authors further described that these changes not only reduce lung volume but also distort it by the infoldings of the vertebral column and changing the angulation of the ribs affecting chest expansion [16].

The compliance of the respiratory system and respiratory muscle strength is related to the Cobb angle [2]. When the angle is <50°, there is minimal effect on respiratory system compliance, however, when Cobb's angle is larger i.e., between 50–100°, the compliance of respiration decreases [2]. The disordered elastic load on the respiratory muscles leads to rapid shallow breathing and low tidal volume and shortened inspiratory time [2, 17]. Other measurements that may assist in predicting the effect of KS on vital capacity include the sagittal diameter of the thoracic cage and the total lung area [17, 18].

The patient had a Cobb angle of 19 degrees of her scoliosis and 73 degrees for her kyphosis, indicating that steroid-induced osteoporosis had a greater impact on the kyphosis than the scoliosis (Figures 4-6). The associated vertebral compression fractures would have certainly aggravated the Cobb angle of the kyphotic component.

Tzelelepis et al. suggest the following indications for NIV in the long term treatment of KS, (i) symptoms such as headache, fatigue or dyspnoea, (ii) signs of Cor Pulmonale such as lower limb oedema, (iii) daytime arterial $pCO_2$ of >45 mm Hg, (iv) <88% nocturnal arterial oxygen saturation [2]. Long term NIV has been shown to reduce the number and duration of hospitalizations and probably increase survival. However, this evidence is not yet supported by randomized controlled studies [2, 19]. In fact, a prospective study done by Gonzalez et al. showed that symptoms in 16 severe kyphoscoliosis patients treated with non-invasive positive pressure ventilation (NIPPV) over a period of 36 months significantly improved with NIV therapy when compared with baseline values. These patients showed a significant improvement in both Pemax and Pimax and nocturnal haemoglobin saturation over the study period, with a better quality of life [20].

Management of patients with kyphoscoliosis and type 2 respiratory failure requires a multidisciplinary approach, which involves various other healthcare professionals. Immunization with Pneumococcal and Influenza vaccines, smoking cessation, keeping an ideal body weight and prompt treatment of acute respiratory infections together with regular exercise are recommended so as to preserve lung function and improve quality of life [2, 9].

Cejudo et al. found that a 10-minute warm-up, 30 minutes of leg exercises on an ergometer cycle and 20 minutes of upper and lower body strength exercises resulted in a decrease in arterial $pCO_2$ and improvement in peripheral muscle strength, dyspnoea and an improvement in the quality of life when compared with controls after 12 weeks [21]. Fuschillo et al. reported that pulmonary rehabilitation improved muscle strength, endurance and quality of life only for a short time, but did not prevent deterioration after one year [22].

Kinnear et al. report that as spinal TB, and poliomyelitis are becoming less frequent, and the numbers of patients with KS affected by these diseases continue to decrease. Nowadays most patients needing NIV at present are older patients with Idiopathic kyphoscoliosis However, mortality after 25 years of NIV was reported to be 40% [23].

## Conclusion

This case clearly outlines the drastic effects severe kyphoscoliosis can have in patients with other underlying common respiratory conditions such as asthma and/or COPD especially when aggravated by steroid induced osteoporosis of the spine . Such an outcome could have been avoided by good patient compliance to inhaler therapy. Domiciliary NIV and 24-hour oxygen therapy via concentrator for respiratory failure offers improvement in symptoms and longer survival together with other supportive measures. Management of such patients requires a multidisciplinary approach across specialities.

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