

Severe Kyphoscoliosis In Pregnancy: A Case Report

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INTRODUCTION

Kyphoscoliosis occurs as a result of disruption of the balance between structural and dynamic components or neuromuscular elements of the spine or the balance and the symmetry of the body as a whole.¹⁻⁵ Kyphoscoliosis is usually idiopathic, involving kyphosis and scoliosis of thoraco-lumbar spine. The incidence of kyphoscoliosis in general population is 1 in 1000 to 1 in 10,000. Mild degrees of kyphoscoliosis are common among pregnant woman. Severe kyphoscoliosis is an uncommon association of pregnancy. Severe kyphoscoliosis could attribute to severe physical disability, and cardio-pulmonary complications.² Pregnancy, superimposed up on this, may have a fatal outcome.

CASE REPORT

A 26-years-old primigravida with short stature presented to outpatient department at 20 weeks of pregnancy. It appeared to be congenital as she had developed a severe spinal deformity (Figure 1) without any history of trauma

or tuberculosis at 1-year of age. During her teenage, she had spontaneous fractures of long bones. On investigating her, Vitamin D levels revealed hypovitaminosis D with high possibility of Osteogenesis Imperfecta. Her Kyphosis Cobb's Angle was 62° and scoliosis Cobb's angle was 92° which is categorized into severe degree of deformity (Figure 2).

She attended outpatient for her regular antenatal visits. After anomaly scan there were no gross anomalies of the fetus and other antenatal profile was within normal limits. As per physician opinion, she started taking vitamin D and calcium

supplements

At 27th week of pregnancy, she was presented to Accident & Emergency unit with breathlessness, her oxygen (O₂) saturation was 78% and was monitored and treated cautiously in high dependency unit with oxygen. Her investigations including chest scan and 2D Echo were within normal limits. She was discharged when she was stable and was advised to take domiciliary oxygen whenever she felt she was breathless.

At 32 weeks, she reported again with breathlessness, swelling of lower limbs and tiredness. Her O₂ saturation



Figure 1 : Patient with spinal deformity

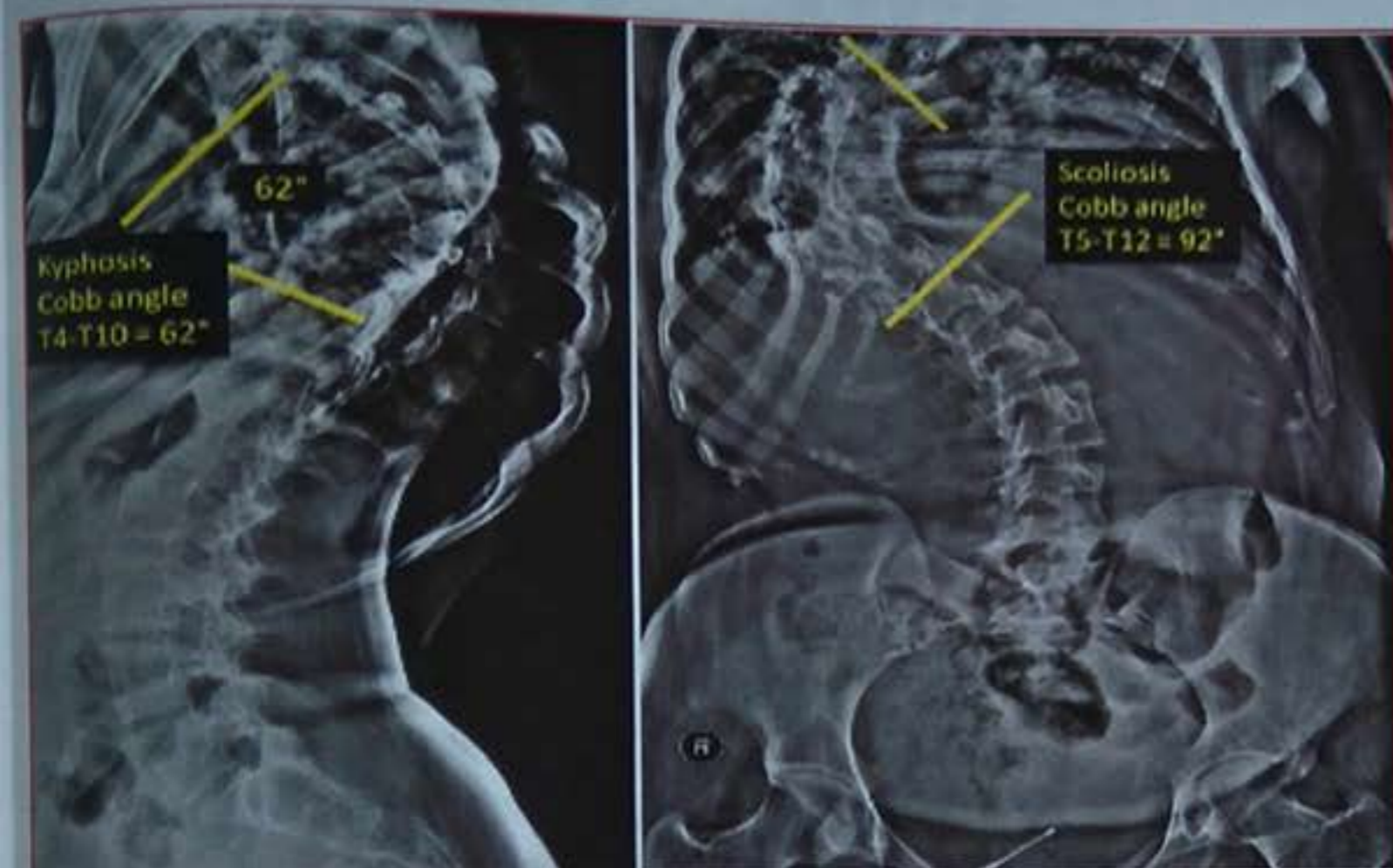


Figure 2

was 82% she had tachycardia, tachypnea and her blood pressure was 140/100 mmHg. Her jugular venous pressure (JVP) was also elevated with significant pedal edema. Further she was shifted to an intensive care unit (ICU). Diagnosis of right heart failure was made. Lung scan revealed mild pleural effusion, 2D Echo reported mild pulmonary artery hypertension with mild diastolic dysfunction. Her ejection fraction (EF) was 52% and she was managed in ICU with diuretics, and nebuliser; subjected to Physician, Intensivist, Pulmonologist and Cardiologist.

On 4th day of her stay in ICU saturation was dropped to 72% and she was put on Bi-Level Positive Air Pressure (BiPAP). CT angiogram was performed which revealed no pulmonary embolism, which showed reassuring remark. Prophylactic low-Molecular-Weight Heparin (LMWH) was started in view of her restricted activity with prolonged stays in ICU. She was in

ICU for 7 days dependent on O₂ with intermittent BiPAP. On Day 8 she was shifted to high dependency unit (HDU) for 10 days, then shifted to ward. She was on O₂ support intermittently.

At 35 weeks of pregnancy, she had another episode of desaturation and required increased oxygen. With deteriorating cardio-pulmonary status, the decision was taken to terminate the pregnancy. Steroid prophylaxis were given to promote lung maturity neonatologist opinion was taken.

With the guidance of senior anaesthetist, an emergency lower segment caesarean section was performed under spinal anaesthesia. Intraoperatively it was difficult to access the lower uterine segment as the patient was in 30 degree propped up position. Also, due to severe kyphoscoliosis there was close approximation of xiphisternum and pubic symphysis thereby reducing the abdominal cavity. Intensive

monitoring of her vitals were done.

All measures were taken to avoid fluid overload. She delivered 2.2 kg male baby with good appearance pulsegrimage (reflex) activity respiration (APGAR). Newborn was examined by a neonatologist and he reported no deformities.

Post operative period, patient was shifted to ICU for observation and was treated with adequate hydration, antibiotics, thromboprophylaxis, analgesics, diuretics and oxygen support. Patient was shifted to ward on postoperative day (POD-5) and was discharged on POD-8 in stable condition. Physician advised her breast feeding and contraception technique and told her to follow-up after a week.

The newborn was screened for vitamin D levels and thyroid-stimulating hormone (TSH) apart from routine newborn screening. Baby was put on calcium and vitamin D supplements by pediatrician team.

DISCUSSION

Severity of kyphoscoliosis is best determined by measuring Cobb's Angle. Kyphoscoliosis is categorized into mild when Cobb's Angle is between 10 to 15 degrees, moderate between 20 to 50° and Severe > 50°. Cobb's angle >60° causes a restrictive defect.¹ With angles 90-100° causes severe restriction. Angle >100° could cause severe deformity and respiratory failure.

Physiological changes in pregnancy, including rise in diaphragm, flaring of ribs in a woman with kyphoscoliosis causes

reduction in the total lung capacity, which includes vital capacity and residual lung volume as there is a relative fixation of the thoracic cage.³

Maternal risk includes cardiac failure, respiratory failure, pulmonary hypertension, increased chances of caesarean section and high maternal morbidity and mortality. With fetus the risks includes intrauterine growth restriction (IUGR), preterm delivery and hypoxic brain damage.

Management in women with severe kyphoscoliosis embarking on pregnancy includes detailed counselling of condition, involving multidisciplinary team. Optimization of her health should be the main focus. Detailed assessment has to be undertaken to rule out other

co-morbidities. Correction of anemia, calcium and vitamin D supplementation if indicated should be advised.

Fetal surveillance to recognize growth restriction, elective preterm delivery is indicated after ensuring fetal lung maturity with steroids being given if preterm delivery is anticipated. More often caesarean section is done as severe kyphoscoliosis is associated with pelvic deformity and maternal cardiopulmonary unstable status.^{3,4} Caesarean section is technically more difficult in patients with severe curves, due to the acute ante flexion of the uterus in the small abdominal cavity resulting from approximation of the xiphisternum and the symphysis pubis the lower uterine segment is inaccessible. Regional anaesthesia is preferred.^{4,5}

Postnatal period may require respiratory function assessment. Long term liaison with pulmonologist, physician should be emphasized.

CONCLUSION

Kyphoscoliosis in pregnancy requires careful evaluation of cardiorespiratory status. Timely admission to hospital, bed rest, maternal and fetal surveillance, multidisciplinary team involvement will reduce cardiorespiratory morbidity and improves perinatal outcome.

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