中文題目：一位嚴重脊柱後側凸病患合併第三型肺高壓經非侵入性呼吸器治療後大幅改善 ──一病例報告

英文題目：A dramatic improvement in group 3 pulmonary hypertension by non-invasive ventilation in a severe kyphoscoliosis patient: A case report

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Background

Treatment of associated condition is the principle in all patients with group 3 pulmonary hypertension. To our knowledge, severe kyphoscoliosis related restrictive lung disease and pulmonary hypertension is relative rare, thus adequate treatment for this condition was uncertain.

Case presentation

We reported a case of 64 year-old female with a history of infantile kyphoscoliosis, and who presented to cardiovascular department due to progressive dyspnea. Severe pulmonary hypertension was diagnosed by transthoracic cardiac sonography (Initial right ventricle systolic pressure: 90.75 mmHg) and right heart catheterization (pulmonary artery systolic pressure was 78 mmHg). Blood gas analysis revealed hypoxemia and hypercapnia. Chest x-ray and chest tomography found severe kyphoscoliosis without active or chronic lung lesions. Chest tomography angiography showed no pulmonary embolism. The lung ventilation/perfusion scan showed no definite evidence of segmental V/Q mismatch involving both lungs. Pulmonary function tests showed FEV1/FVC: 72.74%, FEV1: 0.38L (FEV1%: 48.8%), TLC:1.17L (47.1% predicted), VC: 0.44L (41.2% predicted), and which were compatible with restrictive lung disease. Hence, Non-invasive positive-pressure ventilation (NPPV) without oxygen therapy was applied. Follow-up cardiac sonography revealed dramatic improvement of pulmonary hypertension, and kept steady condition for over 6 years (Initial RVSP: 90.75 mmHg, 3-months follow-up RVSP 37.25 mmHg, 20-months follow-up 46.72 mmHg, 52-months follow-up RVSP: 42.95 mmHg).

Conclusion

In conclusion, even a severe group 3 pulmonary hypertension could be corrected by non-invasive ventilation in patient of kyphoscoliosis.