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Respiratory failure and pulmonary hypertension associated with Klippel-Feil syndrome

Kazuyoshi SUGA,* Kazumi MOTOYAMA,* Akiko HARA,* Norihiko KUME,* Naofumi MATSUNAGA,* Ryosuke KAMETANI** and Masutoku MATSUZAKI**

*Departments of *Radiology and **Second Internal Medicine, Yamaguchi University School of Medicine*

A 28-year-old woman with a deformed thorax and kyphoscoliosis associated with Klippel-Feil syndrome developed respiratory failure with pulmonary hypertension. Pulmonary ^{133}Xe ventilation and ^{99m}Tc-MAA perfusion scintigraphies showed maldistributions of lung ventilation and perfusion, and noticeably delayed ^{133}Xe washout from the lungs. Dynamic breathing MR imaging showed poor and/or asynchronous respiratory movements of the chest wall and diaphragm. These findings indicate that the perfusion-ventilation imbalance, the decreased ventilatory turnover, and expiratory flow from the alveolar space partly derived from the impaired respiratory mechanics may be responsible for the respiratory complications in this patient.

Key words: radionuclide study, Klippel-Feil syndrome, chest deformity, scoliosis, respiratory mechanics, ventilation