Straight Back Syndrome and Respiratory Failure

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**Definition of Straight Back Syndrome**

What is straight back syndrome? As the name suggests, it is characterized by straightness of the backbone. This syndrome was first proposed by Rawlings in 1960 in the Am J Cardiol. According to his paper, 1 straight back syndrome is the absence of normal dorsal curvature (physiological kyphosis) in the thoracic part of the spine, resulting in the reduced anteroposterior diameter of the thorax (the space formed by the ribs, thoracic vertebrae, and diaphragm) and presenting symptoms such as palpitation, chest pain, and shortness of breath, and abnormal clinical findings such as cardiac murmurs and radiographic cardiomegaly.

**Diagnosis of Straight Back Syndrome**

Diagnosis of straight back syndrome is relatively simple. It only requires frontal and lateral chest radiographs, based on which the physician examines the condition of the thoracic spine (whether it is straight or curved dorsally) and the size of the thorax. However, there are 2 sets of diagnostic criteria, one formulated by Davies et al. and the other by De Leon et al. 2,3

In the criteria by Davies et al., we use the lateral chest radiograph and measure the distance $a$ from the middle of the anterior border of T8 to the line connecting T4 (top of the anterior border) and T12 (bottom of the anterior border). Straight back syndrome is diagnosed when this distance $a$ is smaller than 1.2 cm (Fig. 1). This method focuses on the straightness of the thoracic spine.

On the other hand, in the diagnostic criteria by De Leon et al., the anteroposterior diameter $a$ is defined as the distance from the anterior border of T8 to the posterior border of the sternum on the lateral radiograph, and the lateral diameter $b$ is defined at the level of the diaphragm on the frontal radiograph. Straight back syndrome is diagnosed when $a/b$ is 1/3 or less (Fig. 2). This method is considered to detect the reduced anteroposterior diameter of the thorax.

**Relationship between Straight Back Syndrome and Respiratory Failure**

A case of straight back syndrome developing respiratory failure

The literature (Ref. 4) states that patients with straight back syndrome rarely bear the risk of respiratory failure. However, the possibility of respiratory failure is not zero, as my colleagues experienced 3 patients diagnosed with straight back syndrome who developed respiratory failure. Here we briefly look at one of the 3 cases experienced by my colleagues.

The patient was a 52-year-old female with a height of 156 cm and weighing 38.9 kg. The chief complaint was breathing difficulty.
The most remarkable feature of this patient was her slender frame. The physician diagnosed straight back syndrome according to the above diagnostic criteria, and noted the presence of right scoliosis. Dyspnea began when she was about 47 years old, and the patient was receiving medication including expectorants. Elevation of arterial pCO₂ was noted, and this probably was the cause of headache in the early morning. The patient developed pneumonia when she was 48 years old and 50 years old.

When the patient was hospitalized at the age of 52, pulmonary function tests showed that the %VC was 44.1% and the one-second forced expiratory volume rate (FEV1.0%) was 93.3%. Thus, she showed signs of restrictive ventilatory impairment similar to that in other types of thoracic cage abnormalities. Arterial oxygen partial pressure (PaO₂) was 65.1 Torr. Although this did not meet the criterion for chronic respiratory failure (PaO₂ of 60 Torr or less; see below), the decrease in PaO₂ was somewhat abnormal and PaCO₂ was abnormally high at 68.5 Torr. PaCO₂ further increased to 77.6 mmHg after walking. The pH was 7.357, indicating slight acidosis. Differing from the type of respiratory failure seen with common thoracic cage abnormalities, this patient showed a condition resembling type-2 respiratory failure, in which the lowering of PaO₂ occurs concomitantly with the elevation of PaCO₂.

Reminiscent of the sleep apnea syndrome, oxygen saturation measured with a pulse oximeter during sleep was as low as 85% on average and 59% at the minimum. ECG in II, III, and aVF showed ST depression, negative T, and incomplete right bundle branch block.

Echocardiography, lung perfusion scintigraphy, and Ga scintigraphy showed no abnormality.

Based on these observations, this patient with a condition seemingly (though not exactly) consistent with type-2 respiratory failure was considered to have developed this condition as a consequence of straight back syndrome. The deformation of the spine due to this syndrome, as well as right scoliosis and the small size of the thorax, indirectly affected respiratory function over a long period and caused respiratory insufficiency probably in the form of alveolar ventilation (VA) impairment, which often develops to type-2 respiratory failure. The possibility that other respiratory disorders or hidden heart disease might be the cause respiratory failure, however, could not be ruled out.

Features of respiratory insufficiency in straight back syndrome

Unfortunately, the data from our respiratory function laboratory for the previous years were found to include no information concerning
respiratory insufficiency in straight back syndrome. Probably because we did not realize the need, we have not performed respiratory function tests in patients with this syndrome.

We apologize for not being able to provide our own data, but the literature (Ref. 5) states that no significant pulmonary insufficiency was observed, although the total lung capacity (TLC) was somewhat low. We can safely say that the absence of physiological kyphosis in itself does not affect the ventilatory movement of the thoracic cage, despite the potentially reduced size of the lungs. This means that, unlike other types of thoracic cage abnormalities, this syndrome does not imply the risk of respiratory failure. Some patients, however, may actually develop respiratory failure. In such cases, impairment of ventilatory function is considered to result from the small size of the lungs and the abnormality of the thoracic cage containing the lungs.

Patients with common types of thoracic cage abnormalities other than straight back syndrome, such as funnel chest, usually show respiratory function test results indicating restrictive ventilatory impairment characterized by reduced vital capacity. While there are 2 types of respiratory failure, patients with chest deformation are more likely to develop type-1 respiratory failure than the other type. In type 1 respiratory failure, \( \text{PaO}_2 \) decreases, but \( \text{PaCO}_2 \) does not increase.

**Features of cardiac insufficiency in straight back syndrome**

Because of the reduced anteroposterior diameter of the thorax, straight back syndrome is more likely to affect the heart than to cause respiratory insufficiency. This section outlines the effect on the heart.

The cardiologists in our hospital have occasionally encountered cases suspected of having straight back syndrome. According to their accounts and the literature (Ref. 4), frontal chest radiographs of such patients usually show a cardiothoracic ratio (CTR) of 40% or less. The reduced anteroposterior diameter of the thorax often results in protrusion of the left second arc of the heart shadow due to compression of the heart and great vessels, as well as flattening of the heart shadow in about half of all cases. The lateral chest radiograph characteristically shows straight thoracic spine, shortening of the anteroposterior diameter of the thorax, and forward displacement of the heart shadow, often described as so-called small heart syndrome.

ECG is normal in most cases, although right axis deviation, rSr in VI, and small terminal r in aVR have been reported.

**Straight back syndrome and mitral valve prolapse**

However, we need to summarize certain facts about mitral valve prolapse, as it is detected by echocardiography in about half of all cases.

When mitral valve prolapse is detected by echocardiography in straight back syndrome, the anterior cusp of the mitral valve is displaced along the posterior cusp in such a manner that it pushes the posterior cusp slightly toward the left ventricle. The sail-like part of the valve leaflet does not protrude toward the left atrium, and prolapse is localized to the area inside of the center of the anterior cusp.\(^6\)

The mechanism for mitral valve prolapse in these cases has been reported to involve the twisting of the mitral valve during the contraction of the left ventricle, which has been deformed to an oval shape as a result of the reduced anteroposterior diameter of the thorax. The twisting occurs because the anterolateral papillary muscle mainly moves from the lateral to the medial direction, while the posteromedial papillary muscle mainly moves from the posterior to the anterior direction during contraction. When mitral valve prolapse is present, late systolic murmur can be heard at the apex of the heart.

The mitral valve prolapse syndrome can be primary, secondary, or functional. Although the prognosis is generally good, precautions should be taken to prevent the possibility of sudden death.

**Etiology and Prognosis of Straight Back Syndrome**

As for the etiology of straight back syndrome, Bon Tempo et al.\(^7\) reported a theory based on abnormality in the embryonic period. However, because the manifestations of mitral valve prolapse may lessen or disappear with the postnatal growth of the thoracic cage, recent discussion tends to support the importance of a postnatal acquired condition.

The prognosis of straight back syndrome is
usually good, even if complicated with mitral valve prolapse. While specific treatment is generally not required, rare cases developing chronic respiratory failure require treatment for this condition.

Issues requiring future investigation include changes with the growth of the thoracic cage and the positioning of this syndrome as a cause of arrhythmias.

Classification of Respiratory Failure

To provide additional information, this section discusses some facts about respiratory failure. The concept of respiratory failure that is the most widely accepted at present characterizes this condition by the abnormal levels of arterial blood gases, in particular O₂ and CO₂, resulting in the inability of the body to function normally.

However, there is some difficulty with this concept in that arterial blood gases simply reflect the overall functional impairment of the lungs as the gas exchange apparatus, but they do not necessarily detect the condition hindering normal functioning of the body.

Because the ability of the body to function normally is reflected in the gas exchange function at the tissue level, we should measure the mixed venous blood oxygen partial pressure (PvO₂) in the capillary bed of the lungs. While PvO₂ can be measured using a Swan-Ganz catheter, this method is not practical, and the presence of respiratory failure is usually diagnosed based on the analysis of arterial blood gases.

The diagnostic criteria for respiratory failure based on arterial blood gases have been formulated by Filley et al. and Campbell et al. While these criteria developed in other countries are also widely used in Japan, in 1978 the Respiratory Failure Study Team of the Ministry of Health and Welfare proposed the diagnostic criteria for respiratory failure based on arterial blood. The Japanese criteria have also been used in clinical practice recently.⁸

The Japanese diagnostic criteria define respiratory failure as an abnormal condition causing respiratory impairment presenting a PaO₂ (arterial O₂ partial pressure) of 60 Torr or less, or equivalent respiratory impairment. In addition, the criteria classify respiratory failure into type 1, which shows normal PaCO₂ (arterial CO₂ partial pressure), and type 2, which shows abnormal PaCO₂ exceeding 45 Torr. Furthermore, respiratory failure may sometimes be classified into acute respiratory failure and chronic respiratory failure. It seems that acute respiratory failure tends to have causes other than respiratory diseases, while chronic respiratory failure is more likely to be caused by respiratory diseases.

Finally, let us reemphasize the above-mentioned fact that the diagnostic criteria for respiratory failure are based on arterial blood gases, and a PaO₂ (arterial O₂ partial pressure) of 60 Torr or less and a PaCO₂ (arterial CO₂ partial pressure) of 45 Torr or more are important markers.

Because straight back syndrome is asymptomatic in many cases, it is likely to be overlooked. Patients with this syndrome are often diagnosed by general practitioners. When a slender person (particularly a slender female) presents with indefinite complaints such as palpitation, chest pain, and shortness of breath, the physician is recommended to consider the possibility of straight back syndrome even in the absence of specific symptoms. Although we have not confirmed the prevalence in the literature, it is considered to be high.

References