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PULMONARY FUNCTIONS IN PATIENTS WITH IDOPATHIC SCOLIOSIS

İDYOPATİK SKOLYOZ TANILI HASTALARDA SOLUNUM FONKSİYONLARI

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SUMMARY

Idiopathic scoliosis is most common during periods of rapid somativ growth. It is caused by the lateral displacement and rotation of vertebral bodies and has many effects on respiratory function. Scoliosis results in a restrictive lung disease with a multifactorial decrease in lung volumes, displaces the intrathoracic organs, impedes on the movement of ribs and affects the mechanics of the respiratory muscles. Scoliosis decreases the chest wall as well as the lung compliance and results in increased work of breathing at rest, during exercise and sleep. Pulmonary hypertension and respiratory failure may develop in severe disease. In this review the epidemiological and anatomical aspects of idiopathic scoliosis are noted, the pathophysiology and effects of idiopathic scoliosis on respiratory function are described, the pulmonary function testing including lung volumes, respiratory flow rates and airway resistance, chest wall movements, regional ventilation and perfusion studies are presented. Preoperative pulmonary function testing required, as well as the effects of various surgical approaches on respiratory function are also discussed.

Key words: Pulmonary function, spirometry, idiopathic scoliosis

Level of Evidence: Review article, Level V

ÖZET

İdyopatik skolyoz, omurların yana doğru eğilmesi ve kendi etrafında rotasyonundan kaynaklanır. En yaygın olarak büyüme çağında görülmekte olup solunum fonksiyonları üzerinde birçok etkiye sahiptir. Skolyoz intratorasik organların yerini değiştiren, kaburga hareketini engelleyen ve solunum kaslarının mekanik etkilerini etkileyen, kısıtlayıcı-restriktif akciğer hastalığına neden olur. Skolyoz, göğüs duvarını ve akciğer kompliyansını azaltırak dinlenme egzersiz ve uyku esnasında solunum yükünün artması ile sonuçlanır. İlerlemiş hastalıkta pulmoner hipertansiyon ve solunum yetmezliği gelişebilir. Bu derlemede idiyopatik skolyozun solunum fonksiyonları üzerindeki patofizyolojisi ve etkileri tanımlanarak, akciğer volümleri, hava yolu akım ve direnç oranları; göğüs duvarı hareketleri, ventilasyon ve perfüzyon çalışmaları sunulmaktadır. Ameliyat öncesi pulmoner fonksiyon testleri ve çeşitli cerrahi yaklaşımların solunum fonksiyonları üzerindeki etkileri de tartışılmıştır.

Anahtar Kelimeler: Solunum fonksiyonları, spirometry, idiopatik skolyoz

Kanıt Düzeyi: Derleme, Düzey V

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INTRODUCTION

Scoliosis is the most common deformation abnormality of the spine with direct effects on the thoracic cage. Depending on which region of the spine is being affected by the displacement of the vertebrae, the scoliosis is classified as 'thoracic', 'lumbar' or 'thoracolumbar'. Involvement of the thoracic spine (alone or in combination with the lumbar spine) is primary responsible for the respiratory and cardiovascular complications of scoliosis. The prevalence of scoliosis in the general population varying significantly from 0.3 % to 15.3 % [9,13,18-19].

Idiopathic scoliosis is scoliosis for which there is no definite etiology unlike neuromuscular, congenital, or syndromic types and accounts for approximately 85 % of cases. Its diagnosis can be made after exclusion of a primary etiology such as vertebral anomaly, neuromuscular disorder, Marfan syndrome or other disorder.

The effects of scoliosis in the anatomy of the chest are quite complex. Scoliosis can affect pulmonary function in many ways. At an early stage it is usually asymptomatic. Most investigators who have studied the impairment of pulmonary function in scoliosis generally agree that [10] a Cobb angle greater than 90 degrees greatly predisposes to cardiorespiratory failure [16], lung function abnormalities are detectable when a Cobb angle is greater than 50 to 60 degrees [13], lung function abnormalities are mainly of the restrictive type and [9] the duration of scoliosis correlates with the patients degree of disability [15]. Its natural history is associated with curve progression, cardiopulmonary impairment, back pain, cosmetic deformity and neurologic compromise. A great variety on the degree of these manifestations exists, depending on age of onset, genetic background and curve pattern [15].

PATHOPHYSOLOGY AND EFECTS OF IDIOPATHIC SCOLIOSIS ON RESPIRATORY FUNCTION

In the absence of other underlying disorders, mild to moderate scoliosis (Cobb angle less than 70°) actually produces very few respiratory signs and symptoms. Scoliosis has generally been associated with the development of restrictive lung defect, manifested by a decrease in total lung capacity (TLC) on pulmonary function testing. The decrease in lung volume is multifactorial, being determined primarily by the angle of scoliosis (>70), the number of vertebrae involved (seven or more), the cephaladlocation of the curve and the loss of the normal thoracickyphosis [8].

In a recent clinical study, impairment of pulmonary function was seen in more severe cases of spinal deformity, proximally-located curvature and older patients [7].

The decrease in TLC may reflect different pathophysiology depending on the age of the patient at the onset of scoliosis and the chronicity of the problem. Thus, infantile (and possibly juvenile) scoliosis is more likely to be associated with true lung hypoplasia because the thoracic deformity is present during the period of very rapid lung growth and development [3]. In adolescent scoliosis the development, and to a large extent the growth, of the lungs has been completed before the onset of the scoliosis. Thus, the decrease in TLC is more likely to reflect the impaired chest wall mechanics that prevent the normal inflation of the lungs.

True lung hypoplasia due to thoracic deformity during the period of very rapid lung growth and development may be a factor in infantile and possibly juvenile scoliosis [17].

Airway obstruction may occur but is uncommon. Rotation of the chest can produce displacement/rotation of the intrathoracic and/or mainstem bronchi, or compression of a mainstem bronchus against vertebra and mediastinal structures, produce mechanical airway obstruction and reduce expiratory flows and increase airway resistance [5].

PULMONARY FUNCTION TESTING IN IDIOPATHIC SCOLIOSIS

Lung volumes

Restrictive lung disease manifested by a reduction in the TLC is characteristic of severe scoliosis. In such cases simple spirometry may provide a good estimate of the restrictive lung defect because the decrease in the forced vital capacity (FVC) is proportional to the decrease in TLC unless the patient has a mixed restrictive and obstructive defect. Spirometry is much easier to perform in outpatient care settings and is more useful in monitoring the changes in lung function over time, than as the sole means of diagnosing restrictive respiratory disease [1,21]. In patients with moderate to severe scoliosis a negative linear correlation has been established between the magnitude of the curve and FVC.

Patients with increasing coronal and sagittal plane deformities with a high thoracic scoliosis apex are at the highest risk for reduced FVC [4].

A reduction of lung volumes has also been reported in some adolescents with mild scoliosis (e.g. Cobb angle less than 35°), without a clear correlation between the magnitude of the curve and lung volumes. The FVC decreases in proportion to TLC unless there is air-trapping in which case the FVC decreases disproportionately.

Thoracic-dominant scoliosis has an impact on the thoracic cage, there were significant negative correlations between Cobb angle and FVC, and Cobb angle and forced expiratory volume in 1 second (FEV1) values in scoliotic patients [6].

Residual volume (RV) remains generally within the predicted values. Due to the relative decrease in TLC, RV/TLC ratio is

increased. Similarly, the functional residual capacity (FRC) is also normal or slightly diminished and the FRC/TLC ratio is increased. If scoliosis progresses to a severe degree, RV declines slightly. The absolute values of anatomic and alveolar dead space are believed to remain normal. If there is atelectasis and/or hypoinflation of the lung the alveolar dead space will be decreased as well. However, the ratio of dead space to tidal volume (V_D/V_T) is increased. This plays a major role in the development of alveolar hypoventilation.

Maximum inspiratory pressure (MIP) has been reported to be decreased. A crude but significant correlation between the decrease in MIP and the fall in FVC has been reported [13-14]. The decrease in MIP is of major importance in the pre-operative evaluation of the patient because a MIP less than 30 cmH_2O increases significantly the possibility of postoperative respiratory failure due to inability to get extubated. Maximum expiratory pressure (MEP) is normal or may be low, probably due to the chest wall deformity that prevents the muscles from contracting effectively and thus they can not generate the maximal pressure.

In general, the effects on lung volumes in idiopathic scoliosis are the result of reduction of chest wall compliance, impaired lung growth and impaired respiratory muscle strength which work at a mechanical disadvantage.

EFFECTS OF SCOLIOSIS ON THE AIRWAY FUNCTION

Airway function in patients with mild-to-moderate scoliosis tends to be normal. In more severe cases, the maximal expiratory flow volume curve assumes a very characteristic tall and narrow pattern with maximal expiratory flow rates that are elevated disproportionately to the lung volume (reflecting the rapid emptying of the lungs). In addition, the inspiratory flow volume curves, instead of having the semi-circular shape seen in a healthy individual, are virtually a mirror image of the expiratory ones (Fig. 1 and Fig. 2).

As the condition worsens, patients may also develop evidence of lower airway obstruction with concavity in the expiratory limb of the flow–volume curve (Fig. 3). The lower airway obstruction is often reversible with bronchodilators, indicating presence of airway hyperresponsiveness. The latter may be the result of chronic airway inflammation secondary to the poor clearance of secretions [2].

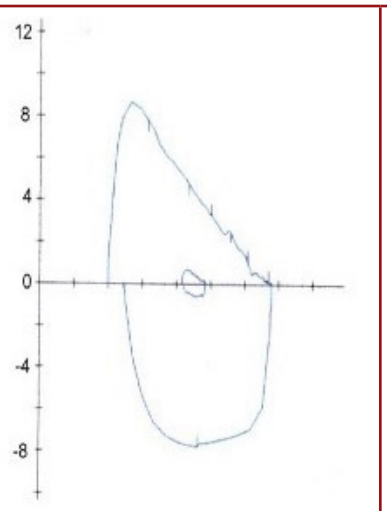

Figure-1. Maximal flow–volume curve (MFVC) in a healthy individual. The expiratory flow–volume curve (above the x-axis) has the typical triangular shape, whereas the inspiratory flow–volume curve (below the x-axis) has a semicircular configuration. Note the size of the tidal flow-volume curve (in the middle) relative to the MFVC. The ratio of tidal volume in relation to the forced vital capacity (VT/FVC) is less than 15%.

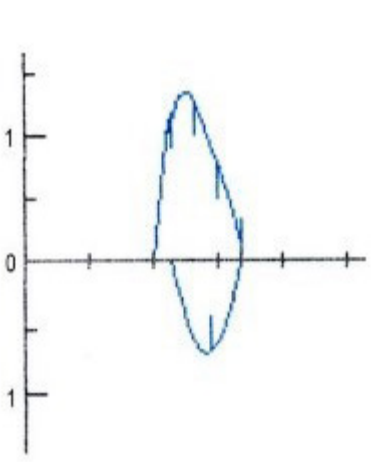

Figure-2. MFVC in a patient scoliosis. The inspiratory curve is a 'mirror' image of the expiratory one, and they both differ in configuration from the normal MFVC. (Forced vital capacity 49% of the predicted normal value; HbSaO2 97 % on room air).

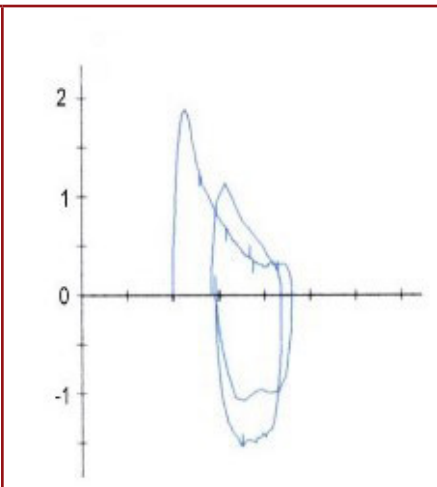

Figure-3. MFVC of a patient with scoliosis showing combined restrictive and obstructive defect. V_T/FVC >60%.

PREOPERATIVE AND POSTOPERATIVE PULMONARY FUNCTION IN IDIOPATHIC SCOLIOSIS

Pulmonary complications are the principal cause of morbidity and mortality in the immediate period following surgery for scoliosis. Preoperative assessment of pulmonary function including TLC and an overnight oximetry should be performed as a guide to prevent postoperative complications.

An FVC less than 40 % of the predicted normal and maximal inspiratory and expiratory pressures of less than 30 cm H2O significantly increase the risk that the patient may not be able to be extubated [(20)].

The presence of preoperative restrictive pulmonary function could be a useful predictor of postoperative pulmonary complications. Moreover, as the FVC ratio worsens, the incidence of postoperative pulmonary complications increases [(11)].

Corrective surgery is used primarily to prevent further progression of the deformity, and secondarily to increase the space available for lung expansion. Indications for scoliosis correction surgery are generally based on severity of curve magnitude and risk of curve progression. In a recent cohort study, scoliosis correction in adolescents was found to increase thoracic volume and is strongly correlated with improved TLC in cases with severe restrictive pulmonary function [(12)].

In summary, idiopathic scoliosis is a common debilitating deformity of the thoracic cage with potentially severe and irreversible effects on lung function. Because the pulmonary manifestations may not become clinically evident until significant or irreversible changes in lung function have already occurred, early recognition of the problem and regular evaluation with pulmonary function testing are advisable.

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