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Spinal bracing for children with neuromuscular scoliosis

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Abstract

Neuromuscular scoliosis (NMS) is a prevalent deformity in patients with neuromuscular disorders (NMDs). It's significantly distinct from idiopathic scoliosis in terms of severity, features, and progression. Treatment of this category is challenging due to the specific natural history and many medical issues related to the deformity. Bracing is required for this population under various clinical conditions to prevent trunk collapse, enhance sitting position, and preserve respiratory function. Its effectiveness in preventing curve progression is still debated, but its value is evident on several levels. The overall goal of this research is to review the role of braces in the management of this uncommon pathology and to raise many essential questions regarding their purpose, effectiveness, various types, and negative impacts of their use.

Keywords: Brace; Neuromuscular diseases; Neuromuscular scoliosis; Duchenne muscular dystrophy; Vital capacity

1. Introduction

Scoliosis is a deformity commonly encountered in many types of NMDs, with an incidence variable according to the severity of the underlying diseases. These disorders generally develop at an earlier age, rapidly progress during growth, and continue to progress even after skeletal maturity. Their management remains a challenging task and varies from conservative approaches to bracing and surgical interventions according to etiology, severity of symptoms, quality of life, and patient/caregiver preference.

The research was conducted by reviewing academic literature on the subject using the above keywords. Six key questions served as the article's main divisions. The first discusses the disease under discussion, its history of evolution, and its pathogenesis. The second explains the role and benefits of the brace. The remaining queries are intended to highlight some of the primary features of this therapeutic approach.

2. Why scoliosis is so common in the NMDs?

NMS is common and severe (60–80%) in individuals with rapidly progressive NMDs, increases with disease duration, and is associated with early loss of walking ability and decreased lung function. However, in slow-progressive NMDs, scoliosis has a relatively low incidence (32%), usually with mild to moderate severity [1]. While NMS can be caused by a number of neuropathic or muscular disorders, the underlying primary etiology is impaired function of the muscular forces acting on the spine, resulting in progressive trunk imbalance. Flaccid, spastic, or dyskinetic paraspinal muscles can occur [2]. The resulting asymmetric forces are insufficient to support spinal curvature and are unable to keep the spine symmetrical against gravity, resulting in an impending imbalance [3].

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3. Why Brace?

Except for steroids for Duchenne muscular dystrophy (DMD), bracing remains the only reliable nonoperative intervention available [2]. However, the decision to brace a patient with NMDs is never easy and requires close collaboration among the multi-disciplinary team. The goal of treatment is to restore and maintain a spine balanced in the coronal and sagittal planes over a level pelvis, to preserve respiratory function, and to halt the progressive loss of functional independence. The brace should be properly made and adjusted to achieve the best results in terms of compliance and comfort while reducing skin irritation. The brace corrects spinal collapse caused by muscle weakness and aims to minimise strain on the fragile ribcage and impaired lungs. It allows to promote growth, correct deformity, and maintain respiratory function until a spinal fusion can be performed [4]. Furthermore, it provides external spinal support, enabling some patients with sensitive skin and curve flexibility to be more functional by improving posture, sitting balance, function, and comfort [5]. Additionally, early treatment requires custom bracing to promote hygiene and delay the onset of cardiopulmonary impairment [6].

4. Side effect?

Moreover, wearing a brace can be uncomfortable and poorly tolerated, leading to decreased brace use, pressure sores, and, in some cases, worsening respiratory problems. In conflict zones, careful fitting and frequent skin checks are required to avoid pressure sores.

5. What effectiveness?

In contrast to the idiopathic scoliosis response to bracing, the data on the effectiveness of bracing in preventing NMS curve progression is still debatable [1]. Although many neuromuscular deformities progress despite bracing, the brace can slow curve progression in patients with smaller curves and muscle hypotonia or mild spasticity [7].

NMS requires lifelong bracing. Early recognition of spinal deformities is critical since treatment plans must be initiated as soon as possible. Whenever a patient's neurological disability is severe, wearing a brace is often perceived as a further impediment to their autonomy and acquisition. In early scoliosis, a brace can easily straighten the curve. But as it progresses, the deformity becomes fixed, and the brace is no longer effective [8].

6. Respiratory level?

The effects of the brace on the respiratory system must be considered in cases where cardiorespiratory function is compromised. They can reduce vital capacity and tidal volume by impeding chest and abdominal expansion [9]. The design of the brace should enable the use of accessory respiratory muscles and maintain chest excursion for adequate ventilation [3]. In severe scoliosis cases with significant respiratory parameters, soft braces may be sufficient to improve posture and balance with better tolerance, although the efficacy of this bracing appears less in the spastic patient [10]. In four studies involving 47 children with scoliosis caused by various NMDs, it appeared that wearing a brace reduced lung function, particularly in cases of DMD and spinal muscular atrophy (SMA). In three studies [11-13], vital capacity decreased by an average of 20% (range: 4–37%) with a spinal brace. One study [14] of eight severe SMA cases revealed that braces reduced tidal volume, increased respiratory rate, and decreased lung compliance.

7. What brace for which patient?

Various braces are used to manage NMS :

Anti-collapse braces, such as the TLSO (thoracolumbosacral orthosis), are used after scoliosis has been structured to support the trunk's functional position, allowing the upper extremities to be used for purposeful activity. They may also be prescribed to prevent collapse, which can limit spontaneous ventilation, cause respiratory infections, increase gastroesophageal reflux disease, and perpetuate an asymmetric posture that promotes the appearance of retractions [15]. The Milwaukee brace is seldom used now for ambulatory patients with stable trunk control [7]. However, it’s not tolerated in advanced cases because of the difficulties in maintaining the balance of the trunk. Some reports suggest a significant impairment of respiration in the brace [14].

When severe deformity is present and the child is too young for surgery, a Garches brace is used to slow progression while awaiting arthrodesis. It's beneficial for patients with respiratory failure and moderate spinal curvature, as it provides good support without straining the rib cage [15]. This brace allows for transfer and seating posture in

hypotonic cases and has a multi-modular component for setting adjustments, with the added benefit of cervical traction and head support for those with severe neck weakness [16]. More conventional braces, such as the Cheneau brace or the hypercorrective nighttime brace, may benefit some patients with disorders of the central nervous system (cerebral palsy, cerebellar ataxia), peripheral nervous systems (neuropathies), or muscular disorders with mild deficits. Likewise, spinal bracing does not work well for certain neurological disorders, such as dystonia [17]

8. Conclusion

Treatment with braces is less effective in neuromuscular patients than in idiopathic scoliosis patients; Nevertheless, this therapeutic approach is able to prevent the collapse of the trunk, improve the sitting position and preserve respiratory function as long as possible.

Compliance with ethical standards

Disclosure of conflict of interest

Authors declare no conflict of interest.

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