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Mini-symposium: Chest Wall Disease

Scoliosis and the Impact in Neuromuscular Disease

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EDUCATIONAL AIMS

- Understand the differential pathophysiology of spinal muscular atrophy and Duchenne muscular dystrophy and how each can lead to scoliosis.
- Understand the considerations involved for when in the progression of scoliosis to intervene.
- Understand the options available for surgical intervention.

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SUMMARY

Scoliosis can alter respiratory mechanics by changing the orientation of the muscles and joints of the respiratory system and in severe forms can put a patient at risk of severe respiratory morbidity or respiratory failure. However, perhaps the most important factor in determining the pulmonary morbidity in scoliosis is the balance between the "load" or altered respiratory mechanics and the "pump" or the respiratory muscle strength. Therefore, scoliosis in patients with neuromuscular disease will both lead to increased "load" and a weakened "pump", an exceptionally unfortunate combination. While progressive neuromuscular disease by its nature does not respond favorably to attempts to improve respiratory muscle strength, the natural approach of early proactive management of the "load" and in the case of scoliosis a variety of different strategies have been tried with variable short term and long term results. Figuring this out requires both an understanding of the underlying pathophysiology of a particular neuromuscular condition and the available options for and timing of surgical intervention.

INTRODUCTION

Patients with progressive hypotonic neuromuscular disease (NMDz) such as spinal muscular atrophy (SMA) and Duchenne muscular dystrophy (DMD) face a gradual decline with increasing respiratory morbidity progressive to the point of respiratory failure. There is a broad range in the rates of decline of the different types of NMDz, but the end point of respiratory failure is often the same. Unfortunately, what is also the same is the current absence of effective treatment options for each of the conditions to prevent the neuromuscular disease that drives this decline. Therefore, the historic and current focus to NMDz treatment is symptomatic and, to the extent possible, preventative therapy.

While the focus of therapy in NMDz is in airway clearance and supporting mechanical ventilation using a variety of proven therapies, there has been a great deal of work over the years to support the respiratory system by treating the progressive scoliosis that occurs in NMDz. Scoliosis in patients with NMDz is different in many ways from that in patients with intact muscle function.

First, the defect in patients with NMDz is based on weakness of the entire muscular component of the thorax, while in congenital scoliosis the scoliosis is typically caused by a significant skeletal defect or in idiopathic scoliosis by an asymmetry in the peri-spinal ligamentous and muscular support of the thorax. Second, because of the diffuse respiratory muscle weakness in NMDz there is less potential to resist the skeletal imbalance that occurs with progressive scoliosis, and as a result it can progress more rapidly. Third, the altered respiratory mechanics produced from scoliosis puts a burden on patients with NMDz that can more

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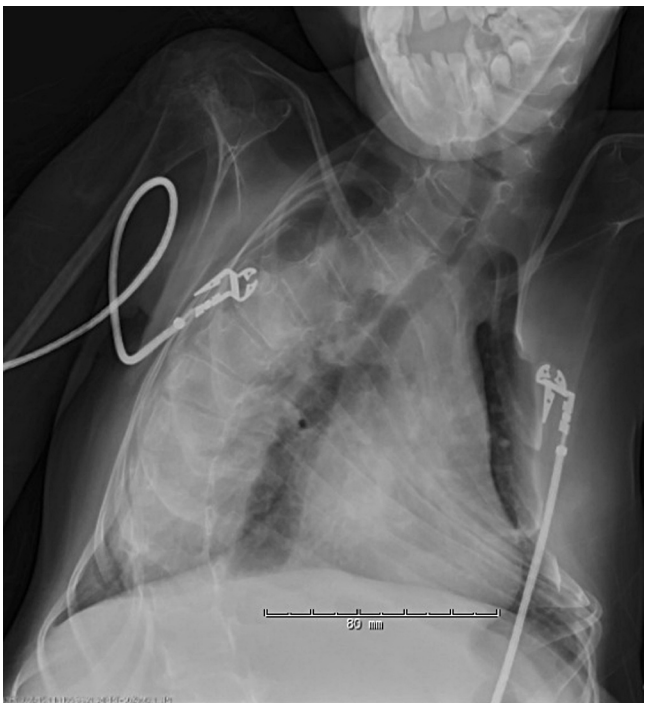

Figure 1. 18 year old female with SMA-2 and uncorrected scoliosis demonstrating severe unilateral rib cage collapse in the convex chest.

easily overwhelm their respiratory muscle function and lead to respiratory failure than in patients with scoliosis and intact muscle strength.

This manuscript will explore the unique features of scoliosis in NMDz and the different ways that it develops in conditions with different pathophysiology, SMA and DMD.

PATHOPHYSIOLOGY OF SCOLIOSIS IN NEUROMUSCULAR DISEASE

In very general terms, spinal muscular atrophy causes more prominent weakness in the muscles of the chest wall than diaphragm, while in DMD the diaphragm is typically weaker with relative preservation of the chest wall muscles [1]. In SMA the

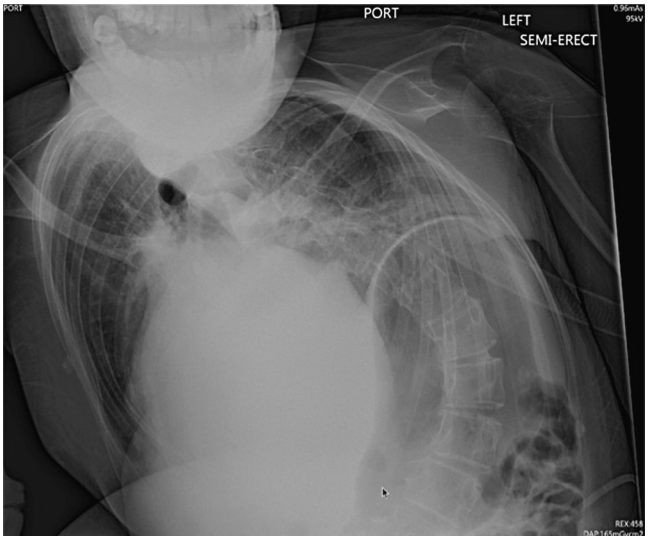

Figure 2. 15 year old male with DMD and uncorrected scoliosis and upward diaphragm displacement.

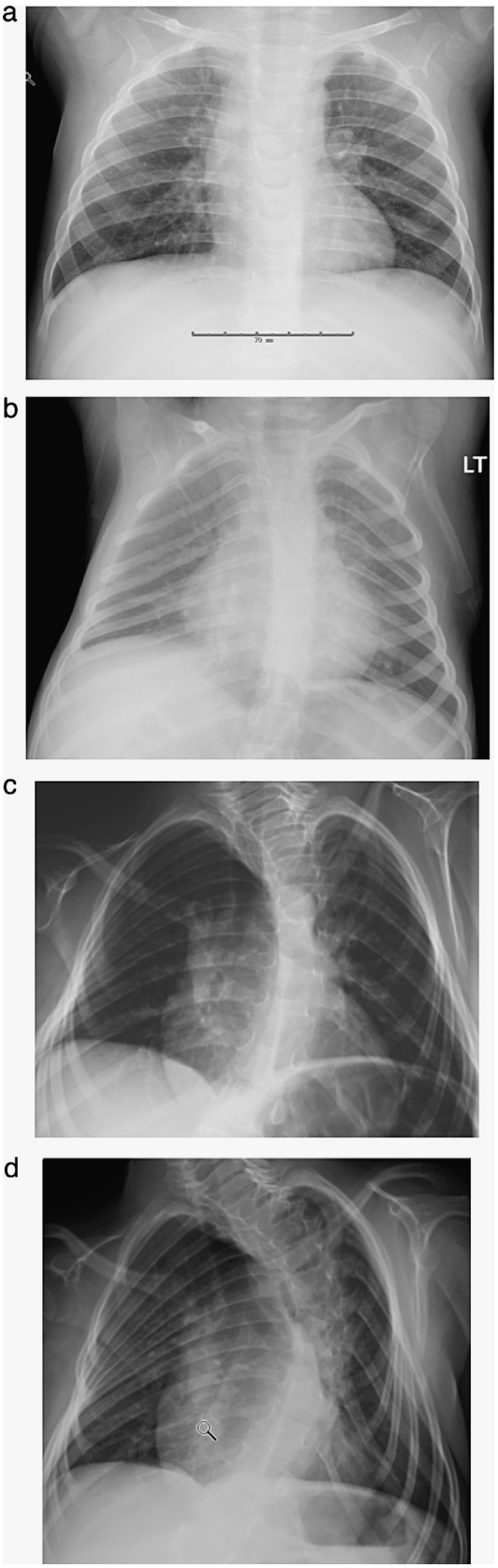

Figure 3. AP chest radiographs in a patient with SMA-2 with progressive scoliosis at a) 10 months of age; b) 3 years of age; c) 5 years of age; and d) 6.5 years of age.