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Review Article

A Review of Surgical Indications for Scoliosis in Duchenne Muscular Dystrophy

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Abstract

The purpose of this article is to review the literature related to indications for scoliosis surgery in patients with Duchenne Muscular Dystrophy. Although the physiology behind spinal deformities in Duchenne Muscular Dystrophy is still unclear, the most recent theory supports the idea of progressive loss of ambulation, poor mobility, and muscle weakness, all leading up to a collapse of the spine. Scoliosis treatment varies from more conservative options, such as physiotherapy and long term glucocorticoids, to surgical intervention of spinal fusion. Duchenne Muscular Dystrophy is a degenerative generalized condition with complications that affect not only the musculoskeletal system, but respiratory and cardiovascular as well. Surgical intervention is essential for improvement of overall quality of life, especially in patients with rapidly declining forced vital capacity, heart failure with left ventricular ejection fraction less than 55% and increasing Cobb angle of more than 35 degrees-or 20 degrees in patients at high risk for rapid deterioration.

Keywords: Duchenne Muscular Dystrophy (DMD); Scoliosis; Spine; Surgery; Indications; Neuromuscular

Abbreviations

DMD: Duchenne Muscular Dystrophy; FVC: Forced Vital Capacity

Introduction

Duchenne Muscular Dystrophy (DMD) is a genetic disease that affects one in 3600 - 6000 live male births [1]. Spinal deformities are commonly observed in DMD, with scoliosis accounting for up to 90% of them [2,3]. In degenerative muscle disorders, scoliosis becomes problematic when the lung function is restricted both by spinal curvature and muscular weakness, affecting the respiratory muscles. In contrast to idiopathic scoliosis there is a significant decrease of vital capacity even in scoliosis with only small curvature [4]. To this day, recommendations of spinal surgery for patients with DMD and scoliosis have been deemed uncertain due to the lack of adequate evidence and scientific support [5].

Scoliosis in Duchenne

DMD is a rare disorder occurring due to the mutations in the dystrophin gene leading to an absence of or a defect in the protein dystrophin [6]. This protein is a part of a complex that works to strengthen muscle fibers and protect them from injury. With its absence, a patient is likely to develop progressive muscle degeneration leading to loss of independent ambulation by the age of 13 years [1]. The type of mutation will dictate the variability of phenotypic expression of this disease.

Majority of the patients are diagnosed with DMD around five years of age. Some of the first signs are noticed by the family members due to the patients' physical abilities differing from that of their peers. The child will demonstrate a delay in achieving certain gross motor milestones, such as not being able to run or jump, and

50% start to walk after the age of 18 months. Respiratory, cardiac, and orthopedic complications commonly appear throughout the years. If left untreated, muscle strength deteriorates, requiring the use of a wheelchair before or during adolescence and ultimately causing mortality at a mean age 19 years [1].

Focusing on musculoskeletal manifestations of DMD, it's widely accepted that poor mobility and continuous muscle weakness alter structures of the trunk leading to progressive collapsing scoliosis. Evolution of the spine deformity in DMD patients is rapid, with an increase in angulation between 16 and 24 degrees per year, fastest being during the adolescent growth spurt [7]. Scoliosis advances from a mean value of about 10 degrees for children under 12 years old to a mean value of approximately 75 degrees by the time they are 16 to 18 years of age [8]. Unexpectedly, DMD patients tend to develop thoracolumbar kyphosis as opposed to the lordosis normally seen in idiopathic scoliosis [7]. According to the series of studies, the direction of the curve is determined by uneven contracture of the iliotibial band. Ambulation, or the lack of it, is a highly significant predictor of the magnitude of the curve [9]. The relationship between ambulation status and scoliosis is reported in one study. It is noted that both wheelchair dependency and spine deformities are age-related phenomena in DMD patients, and the correlation between the two becomes significant 3.5 years after loss of ambulation [10].

Complications of scoliosis in Duchenne

DMD is a rapidly progressive disease leading to the development of scoliosis due to a steadily increasing muscle weakness. Malalignment of the pelvis can generate trouble sitting up along with the difficulties in trunk and head control [4]. Eventually, respiratory muscles become affected as well, leading to pulmonary impairment and circulatory insufficiency [4]. One study reports the rate of decline of normal forced vital capacity (FVC) in 68 patients with DMD, stating that the percentage of normal FVC diminishes quickly during the adolescent growth spurt, leading to the needs of a reliable test to determine the height in these patients [11]. The age at 35% of normal FVC was roughly 15 years and survival for the 28 patients who died was 3.2 (range 0.2 - 5.7) years. Two groups were asked to undergo pulmonary function tests every 10 - 74 months: one group of 21 DMD patients after they underwent spinal stabilization compared with the second group of 46 unfused scoliotic DMD patients. No difference was found in the rate of deterioration of the percentage of normal FVC [11].

Nonoperative management of scoliosis in Duchenne

The mainstay of treatment of DMD consists of physiotherapy and glucocorticoids. Previous studies reveal outstanding results in the use of long-term glucocorticoid therapy by showing its benefits in loss of ambulation at a later age, preserved upper limb and respiratory function, and avoidance of scoliosis surgery [12]. A non-randomized comparative study matched 54 ambulatory males with DMD according to their age and pulmonary function. 30 of the patients were given glucocorticoid deflazacort, while the other 24 received no treatment. The patients were followed for 15 years. Of the patients that survived, 20% in the glucocorticoid treatment group and 92% in the non-treatment group developed scoliosis and underwent spinal surgery. None of the patients in the glucocorticoid group developed scoliosis after ten years of Deflazacort treatment [13]. Just as any other drug, Deflazacort has its unpleasant side effects, such as increased risk of vertebral and lower limb fractures, but the overall impact on quality of life appears positive including the improvement of cardiac function and prolonged ability to walk [14,15].

Another non-surgical treatment option for DMD patients is a thoraco-lumbo-sacral orthosis (TLSO), but there is weak evidence that these braces do not prevent and only minimally delay the onset of scoliosis [16]. Braces may be effective in other forms of muscular dystrophy, but not in DMD [17]. This alternative is impermanent and shows limited effectiveness, it should be reserved for patients who reject an operation or those with inoperable complications due to their general condition of health [16].

Indications for scoliosis surgery in Duchenne

In order to prevent progression of spinal deformities in DMD patients, many have been encouraged to undergo early surgical intervention for spinal fusion and instrumentation to minimize and balance scoliosis [18]. Surgery yields improvement in sitting position and comfort, prevention of vertebral fractures, and reduction in rate of respiratory compromise [7]. Some believe that the correction of the spinal deformity correlates to the improvement of lung function in patients with severe scoliosis secondary to DMD [19]. According to a study performed in 2010, the mean rate of FVC decline in the participating patients after surgery is 3.6% per year. Boys, as well as their parents, notice balance improvement and overall quality of life, even in those with high risk for major com-

plications [20]. When compared to non-surgical groups, the ones who underwent spinal fusion show improved function and decline in the rate of deterioration of FVC. Decrease in muscle power and FVC occurs in both groups, but the deterioration is significantly slower in those after surgical intervention. Even though favorable outcomes are noted regarding the respiratory function, it is still unclear whether or not spinal fusion has any impact on the life expectancy of the DMD patients [21].

The main challenge surgeons face in surgically treating scoliosis in DMD is deciding when to intervene in order to obtain the best possible outcome for each patient. Not all of them develop spinal deformities at the same age, nor do they have similar rates of progression [7]. There is a general rule the surgeons follow: surgical intervention is highly advised and acceptable once the curve sizes reach 35 degrees to prevent critical respiratory decline [22]. But in those at high risk of a rapid deterioration, it is proposed that surgery is performed once the curve reaches 20 degrees and beyond.

Throughout the years, the surgical techniques used for scoliosis fixation have evolved, whether it's a different approach or novel equipment. Surgeons must take into consideration the type of metal to use, the limits of proximal and distal fusion, as well as the source and type of bone graft [7].

Conclusion

Duchenne muscular dystrophy is a rare degenerative condition, which significantly impacts the quality and quantity of a patient's life. Scoliosis has become one of the most important orthopedic complications in patients with progressive muscle conditions. The current literature suggests that surgery of spinal deformities in DMD patients has shown a decrease in deterioration in respiratory and cardiovascular function. Therefore, surgical intervention is essential for improvement of overall quality of life, especially in patients with rapidly declining FVC, heart failure with left ventricular ejection fraction less than 55%, and increasing Cobb angle of more than 35 degrees-or 20 degrees in patients at high risk for rapid deterioration.

Conflicts of Interest

We declare that we have no conflicts of interest in the authorship or publication of this article.

Author Contribution

Authors Phong Truong, Kristina Kuklova, and Natalie Brush contributed to the conception or design of the article; drafted or critically revised the article; approved the final version to be published; and agreed to be accountable for all aspects of the article.

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