Scoliosis in Non-Ambulatory Cerebral Palsy: Challenges and Management

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ABSTRACT: In non-ambulatory patients with cerebral palsy, the presence of scoliosis has a major impact on health and quality of life. The aim of this review is to raise awareness of caregivers from various professions to the extent of the problem, to explain the natural history of neuromuscular scoliosis and its pathophysiology, and to describe up-to-date optional conservative and surgical treatments.

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The presence of scoliosis has a major impact on health and quality of life for non-ambulatory patients with cerebral palsy [1]. The estimated rate of scoliosis in institutionalized patients with cerebral palsy (CP) ranges from 61% to 77% [2-5] and approaches 100% in bedridden children [5]. Koop [4] reported scoliosis of more than 40 degrees at maturity in 30% of quadriplegics, 10% of diplegics, and 2% of hemiplegics. The severity of the neurological impairment is apparently the most strongly associated factor with the magnitude of the curvature [5]. Children with a level IV impairment according to the Gross Motor Function Classification System (GMFCS) have a 50% greater risk of moderate or severe scoliosis by 18 years of age than children with GMFCS level I–II [6].

Spinal deformities in non-ambulatory patients with cerebral palsy are common and cause posture related problems such as pain and imbalanced sitting, and pose a significant challenge to patients, families, and caregivers

NATURAL HISTORY

The progression of scoliosis is related to age at curve initiation and the initial Cobb angle [7]. Saito et al. [5] reported that a spinal curve of 40 degrees before 15 years of age carries an 85% risk of spinal curve progression to more than 60 degrees. In adult patients, neuromuscular scoliosis tends to progress in correlation to curve severity [3,5]. Collapsing curves in wheelchair users may cause painful costopelvic impingement leading to sitting intolerance in addition to upper extremity dysfunction and a consequent need for a modified wheelchair to improve support [8]. Cardiopulmonary complications may arise due to the chest distortion associated with spinal deformity, and pressure sores may develop as a result of asymmetrical loading and pelvic decompensation [9].

CURVE PATTERN

The neuromuscular scoliosis that occurs in non-ambulatory patients with CP typically involves the complete spine and creates a long C-shaped curve that includes the sacral vertebrae, often with pelvic obliquity [10]. Although the convexity may be directed to the right or left, Saito et al. [5] found that most thoracic and thoracolumbar curves (75% and 86%, respectively) were to the right, and most lumbar curves (73%) were to the left. Typically, the scoliotic curve is associated with sagittal thoracic kyphosis, although some patients may present with a hyperlordotic deformity [11] with anterior pelvic tilt and hip flexor contractures [9]. Hypolordosis of the lumbar spine was found in relation to the presence of severe hamstring shortening, with a resultant posterior pelvic inclination [12]. This finding may explain the development of the compensatory thoracic kyphosis [9]. The magnitude of the Cobb angle is correlated with the vertebral rotation in the transverse plane [13]. The consequent horizontal pelvic rotation may adversely impact sitting balance and hip stability [14].

PATHOPHYSIOLOGY

The association of scoliosis with spasticity is well documented [2,5], but the pathophysiology underlying the formation of scoliosis is unclear. Muscle weakness, asymmetric tone, and incompetent paraspinal muscle control with resultant truncal imbalance, and poor postural control seem to be etiologies for scoliosis in patients with CP, but supportive data are still lacking [15,16]. Presumably, these mechanisms are responsible for the development of scoliosis in children with all types of neuromuscular conditions [15].

ASSESSMENT

Non-ambulatory patients with CP and scoliosis warrant proper multidisciplinary assessment with consideration of all treatment options. A comprehensive clinical history is essential, starting with the perinatal period, including growth and development parameters, medical diagnoses and treatment, and the achievements of any developmental milestones.

Patients and caregivers should be questioned about pain levels, sleep disorders, self-expression ability, feeding problems, upper extremity function in activities of daily living, and access to rehabilitation programs. All of these factors can affect decision-making. In addition, the medical records should be reviewed for potential risk factors of surgery, namely occurrence of seizures requiring medication, recurrent urinary and pulmonary infections, presence of gastroesophageal reflux, signs of malnutrition, and poor bowel function. These parameters should be thoroughly tested during physical examination.

To assess level of activity and ambulation, range of motion, muscle tone, and muscle power should be measured. Hip flexion contractures and short hamstrings can affect truncal position, and pelvic tilt and rotation in the horizontal plane may arise due to hip instability and dislocations. It is not entirely clear if scoliosis develops before or after hip asymmetry, or if the prevention of hip subluxation or dislocation decreases the frequency or severity of scoliosis. However, windswept deformity, pelvic obliquity, and scoliosis are related and may worsen concomitantly [17-19]. Curve flexibility in both the sagittal and coronal planes can be tested by gentle upright traction under the shoulders [20]. Patients with a severely imbalanced sitting position should be examined for skin sores over prominent areas such as the sacrum, ischium, or greater trochanter, or within skin folds in the flank region [4,16,17]. Systemic function needs to be evaluated, with an emphasis on swallowing test and sputtering during feeding, which may be signs of aspiration risk. Pulmonary function is assessed either directly or by indirect signs of vital capacity (such as crying, laughing, and vocalizations) [21,22]. Albumin levels may serve as a marker of nutritional status, and lymphocyte count as a marker of immune status [23].

Anteroposterior and lateral views of the entire spine should be obtained in the seating and standing positions if possible. Although there is no consensus regarding technique and patient positioning, they should be unique for each patient and determined by the individual's functional ability [16]. Anteroposterior scans provide information on curve type, curve magnitude, spinal balance, spinal rotation, rib deformity, and vertebral wedging [17]. Pelvic obliquity is expressed as the angle between a line drawn perpendicular from the middle of T1 and S1 and the line connecting the top of both iliac crests [24]. Scans made in the supine traction view (gentle manual traction on the legs and counter-traction below the shoulders or with a head halter and pelvic straps) [20] or with side flexion if possible, provide information on spinal flexibility and the measure of potential correction. The lateral view is important for assessing overall sagittal balance.

Although spondylolisthesis has been documented in 21% of cases of ambulatory spastic diplegia [25], there are no reported cases in the non-ambulatory population, so routine assessment in the setting of scoliosis in CP is probably unnecessary [26].

CONSERVATIVE TREATMENT

Seating adaptations may be beneficial for improving trunk balance, maintaining an upright posture, and facilitating patient care and function. Specific measures include molded wheelchair inserts, adjustable head support, chest lateral supports, and shoulder and trunk harnesses and straps. Wheelchair tilt is helpful. In patients with fixed rigid deformities, the back of the seat should be molded. Improved trunk support leads to better head and neck control and better use of the upper extremities [27]. Upright positioning has also been shown to positively affect respiratory parameters [28,29].

Conservative treatment with braces and adjusted trunk supports improve sitting positions but do not prevent curve progression

Spinal bracing has been advocated for the management of neuromuscular scoliosis, with the same benefits as seating adaptations. In general, in spastic patients, soft braces are better tolerated than rigid orthoses for skin integrity [30], with no difference in pulmonary mechanics or gas exchange [31]. Although bracing may slow the deterioration in flexible curves [27,32], there is no evidence that it controls curve progression [33-35]. Moreover, it can considerably reduce forced vital capacity, which may contribute to poor tolerability of the brace, particularly given the already impaired lung function [29]. If the brace is too tight, it can cause skin irritation and respiratory compromise and exacerbate gastroesophageal reflux and feeding/swallowing disorders [9, 27].

SURGICAL TREATMENT

Spinal fusion surgeries are intended to achieve a level pelvis and brace-free balanced sitting, halt curve progression, and alleviate pain. Since they carry significant risks, the specific treatment goals must be well defined, realistic and meet the medical conditions or needs of the individual patient [20]. The natural history of the curve progression should be kept in mind when considering the pros and cons of surgery [1,3]. Retrospective self-report (patient and caregiver) and questionnaire-based surveys of the outcome of spinal fusion in patients with CP reported a significant improvement in health-related quality of life and very high satisfaction rates in terms of improved sit-

ting balance, cosmesis, and quality of life; however, functional improvements were limited [36]. Most parents (95.8%) and caretakers (84.3%) of children with spastic disorders would recommend surgery [37]. A long mean predicted survival of 11.2 years has been reported in children and adolescents with severe spastic CP and neuromuscular scoliosis who underwent surgical correction [38]. Since there was no available control group of severely affected children who did not undergo spinal fusion, the authors could not reach a definitive conclusion regarding the impact of surgical correction on life expectancy in the pediatric age group. The most accurate determinants of survival were the number of postoperative days in the intensive care unit and the presence of excessive thoracic hyperkyphosis [38].

Spinal fusion is at present the only effective treatment to have a positive impact on quality of life and nursing care

In institutionalized mentally retarded patients, the indications for surgery should be carefully evaluated. A comparison of health and nursing issues between operated and non-operated institutionalized patients with CP found no significant difference in pain, pulmonary status, decubitus ulcers, function, or time required for daily care. However, the nurses caring for the patients believed that those who had undergone spinal fusion were more comfortable [39].

SURGICAL COMPLICATIONS

Neuromuscular deformities have the highest surgery-related morbidity (17.9%) and mortality (0.34%) of all types of scoliosis [40]. Complications include implant or bone fixation failure with need for revision surgery, pulmonary, gastrointestinal, and neurologic impairments, poor wound healing, deep wound infection, long hospital stay, and death. With steady improvements in surgical techniques and perioperative management, the risk of major complications has been minimized [20].

CONCLUSIONS

Spinal deformities are common in non-ambulatory patients with CP and proportional to the neurological involvement. They cause posture-related problems of back and rib pain as well as functional problems due to imbalanced sitting, and pose a significant challenge to patients, families, and caregivers. Conservative treatment with braces and adjusted trunk supports improve sitting position but do not prevent curve progression. The only effective treatment at present is spinal arthrodesis, which has been found to prevent deterioration, improve sitting balance, and reduce the spinal curvature with a positive impact on quality of life and nursing care.

A multidisciplinary management approach is needed, including careful preoperative and operative planning on an individual basis. Factors that can impact surgical outcome, such as nutritional status, pulmonary function, the presence of gastrointestinal reflux, bowel motility, seizures, coagulopathies, and current medications must be addressed.

Young patients are at significantly higher risk of severe and often life-threatening perioperative complications, and novel instrumentation techniques have been introduced in recent years to preserve spinal growth in this population. Families and caregivers need to be carefully counseled to ensure that they understand and accept the risks involved and maintain realistic expectations.

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Capsule

MORE model to fight against addiction

The United States is in the midst of an opioid crisis affecting more than 15 million Americans. **LaBar** and co-authors provided neurophysiological evidence for a therapeutic approach to help individuals with opioid use disorder (OUD). They conducted a series of randomized experiments using an approach called Mindfulness-Oriented Recovery Enhancement, or MORE. Hedonic dysregulation in brain reward circuitry has been viewed as a core mechanism of addictive behavior. The MORE model aims to restructure reward responsiveness using mindfulness techniques. MORE may remediate hedonic dysregulation by simultaneously increasing responsiveness to natural rewards and decreasing reactivity to drug-related rewards. The use of MORE and other cognitive training interventions may ultimately serve to turn the tide on addiction in the United States.

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Capsule

Acute flaccid myelitis may be caused by a virus

Ayers et al. examined comprehensive surveillance data for the polio-like illness, acute flaccid myelitis (AFM), in the United States from 2015 to 2017. It can be every mother's worst nightmare. Her lively 2-year-old starts slurring his speech and is unable to move an arm to share their usual high fives. This nightmare became a reality for the parents of several children in Colorado who were diagnosed with AFM, a neurologic condition in people ≤ 21 years of age, characterized by acute onset of flaccid limb weakness with magnetic resonance imaging results showing predominantly gray matter lesions on the spinal cord. These signs differentiate this condition from other forms of paralysis, such as Guillain-Barré syndrome. AFM made national news in 2018, with a total of 228 confirmed cases in 41 U.S. states, and four confirmed cases in 2019. The spike in reported cases in 2018 was so dramatic that the U.S. Centers for Disease Control and Prevention announced they would start tracking AFM cases.

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