Common Complications of Pediatric Neuromuscular Disorders

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KEYWORDS

• Pediatric neuromuscular disorder • Nutrition • Constipation • Scoliosis
• Hip dysplasia • Sleep

KEY POINTS

- Children with pediatric neuromuscular disorders suffer from common complications.
- The complications are related to immobility and weakness.
- Scoliosis, hip dysplasia, and osteoporosis are common musculoskeletal complications.
- Constipation, gastroesophageal reflux, obesity, and malnutrition are common gastrointestinal complications.
- Disordered sleep affects both the children and their caregivers.
- Screening for these common complications can lead to healthier children with pediatric neuromuscular disorders and a resulting higher quality of life.

INTRODUCTION

Nature of the Problem

There are several pediatric neuromuscular disorders (pNMDs), such as cerebral palsy (CP), myelomeningocele, spinal cord injury, chromosomal abnormality, acquired brain injury, acquired and hereditary neuropathy, myopathy, and motor neuron disorders that all share common complications. Immobility and weakness are the primary etiologies for most of these commonly seen conditions. Musculoskeletal complications in pNMDs include hip dysplasia with associated hip subluxation or dislocation, neuromuscular scoliosis, and osteoporosis and resulting fractures. Constipation and gastroesophageal reflux (GER), along with obesity and malnutrition, are commonly

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experienced gastrointestinal (GI)-related complications. Disordered sleep also is frequently observed, and this affects not only the patients but also caregivers.

MUSCULOSKELETAL COMPLICATIONS

Musculoskeletal complications, such as limb contractures, hip dislocation or subluxation, and scoliosis are common in pNMDs (Table 1). They contribute to increased disability due to decreased motor performance, mobility limitations, reduced functional range of motion, loss of function for activities of daily living (ADLs), decreased quality of life (QOL), and increased pain.

Scoliosis in pNMDs leads to multiple problems, including poor sitting balance, difficulty with upright seating and positioning, pain, and preclusion of the ability to sit upright in a wheelchair.[1] Screening for spinal deformities is important because it can have several clinical implications. Unfortunately, spinal deformity is neither preventable nor responsive to nonsurgical modalities such as bracing. Unlike idiopathic scoliosis, neuromuscular scoliosis almost always progresses. Early detection and screening are crucial for proper and ideal management of scoliosis.

The Adam forward-bend test is the primary screening test for neuromuscular scoliosis and should be performed on all patients with pNMDs. The screening examination is performed by having the patient bend forward as far as possible, flexing the cervical and thoracolumbar spine. If a patient is unable to stand, this can be performed in the seated position. Some patients will require postural support if they are unable to sit independently. The patient is viewed from behind focusing on the rib cage. The examiner is looking for one side of the rib cage to be higher than the other next to the vertebral column. The convex side of the scoliosis is the side with the rib hump. In obese patients, smaller curves can be missed, especially in the lower lumbosacral spine.

If a spine curve is detected or the patient's body habitus precludes the test's sensitivity, spinal radiographs should be performed. Anteroposterior (AP) and lateral spinal radiographs with the patient either sitting or standing, based on the individual's function, are generally sufficient. On the AP film, the Cobb angle is measured.[2] Serial measurements should be performed using the same anatomic landmarks to ensure comparable measurements (Fig. 1).

Hip subluxation and dislocation due to hip dysplasia are frequently encountered in children with pNMDs. Hip dysplasia is a condition of the hip that may be present at or shortly after birth with inadequate acetabular formation. At birth, neonates have a shallow acetabulum. As they grow, the acetabulum usually deepens and contours around the femoral head. When infants have decreased muscle tone, strength, and movement, the acetabulum remains shallow due to the reduced force applied to the acetabulum by the femoral head. Hip dysplasia is most commonly seen in pNMDs

Table 1
Incidence of musculoskeletal complications of pediatric neuromuscular disorders

Musculoskeletal Complications	Cerebral Palsy	Myelomeningocele	Duchenne Muscular Dystrophy	Spinal Cord Injury	Charcot-Marie-Tooth	Spinal Muscular Atrophy
Scoliosis, %	38–64	20–94	63–90	100[a]	10	70–100
Hip dysplasia, %	2–60	1–28	35	29–82	6–8	11–38

[a] If injured before adolescent growth spurt.

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