POSNA
The Core Curriculum

Neuromuscular scoliosis

Objectives

1. Define neuromuscular scoliosis.
2. List causes of neuromuscular scoliosis. Include upper motor neuron, lower motor neuron, and myopathic causes.
3. Describe the genesis and development of neuromuscular spinal deformity.
4. Describe the role of pelvic obliquity in neuromuscular scoliosis.
5. Discuss the role of bracing for neuromuscular scoliosis.
6. Describe factors contributing to the higher complication rate following surgery for neuromuscular spine deformity.

Discussion points

1. What sagittal deformities accompany neuromuscular scoliosis? Why?
2. What differences present with a neuropathic etiology as opposed to a myopathic etiology?
3. How can one evaluate nutritional status prior to operative intervention?

Discussion

Neuromuscular spinal deformity comprises some of the most challenging problems encountered in pediatric orthopaedics. The Scoliosis Research Society has classified neuromuscular scoliosis into neuropathic and myopathic causes. Neuropathic lesions include upper motor neuron lesions such as cerebral palsy, Frederich's ataxia, and spinal cord trauma and tumors. Even in this group there is considerable variety in the effect on the spine - cerebral palsy usually includes a spastic component, while spinal cord trauma and tumors may or may not depending on the site and nature of the lesion. Neuropathic causes also include lower motor lesions such as poliomyelitis, and spinal muscular atrophy. Myopathic lesions include arthrogryposis, muscular dystrophy, and myotonia; the spinal deformity patterns from these etiologies also vary considerably. Despite these differences, the common factor is an inability to provide muscular support to the spinal column. Kyphosis commonly accompanies neuromuscular deformity secondary to forward tilt of the head and/or poor head control. Once the curve is present, gravity and posturing provide the impetus for worsening deformity. Age of onset obviously varies with etiology, for example, spinal muscular atrophy produces deformity at a younger age than muscular dystrophy.

It is important to view the sacrum (and the rigidly attached pelvis as the last vertebral segment. The lumbosacral joint can deform in any plane as the rest of the spine, or more proximal deformity can throw the pelvis into an oblique position without lumbosacral deformity. Contrarily, hip contracture can secondarily deform the spine dynamically when the patient attempts to

accommodate the hip deformity while sitting. It is important to carefully assess hip motion and contracture in any patient with neuromuscular spinal deformity. Small amounts of pelvic obliquity (< 10-15 degrees) are compatible with comfortable seating. Larger fixed pelvic obliquities are not, and must be corrected by operative or (if not fixed) wheelchair modifications.

The goal of treatment is a balanced spine and pelvis, generally for the purpose of sitting; although some patients with, for example, arthrogryposis or Frederich's ataxia, can ambulate into adult life. Modalities used to treat neuromuscular scoliosis include wheelchair modifications in addition to the usual bracing and surgery. Bracing has traditionally been felt to be ineffective, although recent reports are more encouraging. It is technically difficult and time consuming, but success has been reported with both cerebral palsy and muscular dystrophy patients. The ultimate role of bracing is unclear.

Non-ambulatory patients with muscular dystrophy with curves over the 20-30 degree range have been regarded as candidates for surgery, the rationale being that the curve will inevitably progress and pulmonary function will decrease. Technical success has been routine in recent reports. Blood loss is higher in children with muscular dystrophy, hypotensive anesthesia has been helpful. Patient satisfaction is generally high after scoliosis correction in the muscular dystrophy patient.

The role of surgery for the cerebral palsy patient has been less well defined. A study of institutionalized patients did not clearly define benefit, although the patients were reported as being more comfortable. Higher complication rates have often been reported, and nutritional assessment has been recommended prior to surgery. An albumin level of >3.5/dL and a total lymphocyte count of >1500/mm3 were regarded as minimum requirements. Another study regarded the severity of overall involvement of the patient, recent medical problems, and severity of curve to be more significant. Both may well be correct, as the most impaired non-ambulatory patients with cerebral palsy develop the most severe curves.

The state of knowledge regarding neuromuscular scoliosis is presently undergoing some revision; this process is dynamic and current literature review is essential. The somewhat large bibliography for this subject reflects the divergent opinions currently presented.

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