Preferences and Attitudes to Management of Equinus in Cerebral Palsy Children: A Survey of Orthopaedic Surgeons

Thesis

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INTRODUCTION

Cerebral palsy (CP) is a leading cause of childhood disability worldwide. Motor dysfunction is a universal feature among CP children, [1, 2] with ankle equinus as one of the extremely prevalent deformities in CP children [3].

Equinus contracture is best remedied by lengthening of triceps surae or gastrocnemius-soleus (gastrocsoleus) complex. Numerous triceps surae lengthening procedures and their modifications are described in the literature [4-10].

They have been classified in accordance with three distinct anatomic zones of triceps surae complex. Zone one comprises the medial and lateral bellies of the gastrocnemius, their tendentious insertions and soleus muscle belly. Zone two comprises the aponeurotic tendons of the gastrocnemius and soleus, and Zone three comprises the Achilles tendon [11].

Generally, the more proximal procedures are more selective and stable but are not well suited for severe deformities where greater degrees of correction at the ankle are required. The distal lengthening procedures are less selective and less stable but allow for greater degrees of correction [6, 12-16].

Crouch gait also known as excessive knee flexion gait Is frequently encountered in ambulatory children with bilateral spastic CP formerly known as spastic diplegia. Loss of plantar-flexion/knee-extension coupling mechanism is a notable risk

factor contributing to the development of crouch gait. Additional risk factors include muscle weakness and spasticity [17, 18].

Neglected crouch gait can lead to a significant functional deterioration through the creation of lever-arm dysfunction and eventual joints contractures and bony deformities [17-20].

On the other hand, imprecise surgical interventions can aggravate the child's gait inefficiency. This is especially true for the management of ankle equinus. For example, overlengthening, or poorly selected triceps surae lengthening procedure (as an unnecessary Achilles tendon can lengthening) result in the development of a calcaneus deformity deterioration of crouch gait with diminished ankle push-off moment.

This is usually referred to as iatrogenic or "surgeon-induced crouch" the pathogenesis of which occurs through insulting the soleus an already overburdened muscle in crouch patients. [17, 18, 21, 22] Likewise, a mistimed -usually early in life- triceps surae lengthening procedure can precipitate deformity [3, 22].

Silfverskiold's test plays a pivotal role in differentiating gastrocnemius-induced contracture from combined gastrocsoleus-induced contracture. Nonetheless, it is commonly overlooked by physicians in the clinical assessment of ankle equinus [23, 24].

Surgeons' preferences and attitudes to the management of pediatric CP hip migration [25] and pediatric fractures [26] have

been shown to exhibit diversity on a national and international scale. Such diversity in the surgeons' viewpoints may impact on the treatment outcome and relevant complication rates. The caregiver's and parent's attitudes are equally important to the health status of CP children. [27]

Surveys are employed to convey opinions, beliefs and current practice held by physicians throughout their practice. Medical surveys are essential tools to uncover gaps between research and practice

The consensus among orthopedic surgeons as to the management of equinus deformity in CP children has not been reported previously despite being a prevalent deformity [25, 26, 28].

AIM OF THE WORK

The goal of this survey study is to examine the orthopedic surgeon's preferences and current practice to the evaluation and surgical management of equinus deformity in children with ambulatory CP.

The specific aim of the study is to identify and analyze variations in current practice and decision making between general orthopedic and pediatric orthopedic surgeons.

Chapter 1

CEREBRAL PALSY

Cerebral palsy (CP) is a term that has been formally defined as a group of permanent disorders of the development of movement and posture, causing activity limitation, which is attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication and behavior, by epilepsy and by secondary musculoskeletal problem [29].

Prevalence estimates range from 1.5-3/1,000 in western countries, with much higher and wider range, 2-10/1,000 live births, in the developing areas [30].

70 to 80 % of cerebral palsy cases are acquired prenatally and from largely unknown causes. Birth complications, including asphyxia or hypoxia are currently estimated to account for about 6 % of patients with congenital cerebral palsy [31].

Neonatal risk factors for cerebral palsy include birth after fewer than 32 weeks' gestation, birth weight of less than (2,500 gm.), intrauterine growth retardation, intracranial hemorrhage, and trauma in about 10 to 20% of patients, Cerebral palsy is acquired postnatally, mainly because of brain damage from bacterial meningitis, viral encephalitis, and hyperbilirubinemia[31].

Diagnosis of Cerebral palsy

The diagnosis begins with a history of gross motor developmental delay in the first year of life. The general medical history should include a review of systems to evaluate for the multiple complications that can occur with cerebral palsy.

Prenatal history:

The prenatal history should include information on the mother's pregnancy, such as prenatal exposure to illicit drugs, toxins, or infections; maternal diabetes; acute maternal illness; trauma; radiation exposure; prenatal care; and fetal movements. A history of early frequent spontaneous abortions, parental consanguinity, and a family history of neurologic disease (eg. hereditary neurodegenerative disease) is also important [32].

Perinatal history:

The perinatal history should include the child's gestational age (ie, degree of prematurity) at birth, presentation of the child and delivery type, birth weight, Apgar score, and complications in the neonatal period (eg, intubation time, presence of intracranial hemorrhage on neonatal ultrasonogram, feeding difficulties, apnea, bradycardia, infection, and hyperbilirubinemia)[33].

Developmental history:[34]

The child's developmental history should review his/her gross motor, fine motor, language, and social milestones from birth until the time of evaluation. Gross motor milestones of concern with cerebral palsy include head control at age 2 months, rolling at age 4 months, sitting at age 6 months, and walking at age 1 year. Infants with cerebral palsy may have significantly delayed gross motor milestones or show an early hand preference when younger than 1.5 years, suggesting the relative weakness of one side (eg. reaching unilaterally).

Observation of slow motor development, abnormal muscle tone, and abnormal posture are common features to the diagnosis of cerebral palsy.

Assessment of persistent infantile reflexes in normal infants, the Moro reflex is rarely present after six months of age, and hand preference rarely develops before 12 months of age.

Cerebral imaging using computed tomography, magnetic resonance imaging, and ultrasound are useful physical diagnostic tools.

Surveillance for associated disabilities such as hearing and vision impairment, seizures, perception problems with touch or pain, and cognitive dysfunction can help complete the clinical assessment and determine the diagnosis.

Classification and Assessment of Cerebral palsy

CP is divided into spastic, dyskinetic, and ataxic subtypes. The spastic subtype is further divided into unilateral type (limb involvement on one side of the body) and a bilateral type (limb involvement on both sides of the body [35].

The Gross Motor Function Classification System (GMFCS)[36]

Is a classification system developed for children with CP. Initially, children with CP were divided into five levels by considering their independency in gross motor functions such as sitting, walking, mobilization and transfer activities and the tools-equipment.

Motor function is classified based on walking ability. Children classified as GMFCS Levels I or II were categorized as 'walks independently', Level III as 'walks with handheld mobility device', Levels IV as 'limited walking ability', and level V as wheel-chair bound

GMFC classification system:

LEVEL I - Walks without Limitations, LEVEL II - Walks with Limitations, LEVEL III - Walks Using a Hand-Held Mobility Device, LEVEL IV - Self-Mobility with Limitations; May Use Powered Mobility, LEVEL V - Transported in a Manual Wheelchair.

Distinctions between Levels I and II

Compared with children and youth in Level I, children and youth in Level II have limitations walking long distances and balancing; may need a hand-held mobility device when first learning to walk; may use wheeled mobility when traveling long distances outdoors and in the community; require the use of a railing to walk up and down stairs; and are not as capable of running and jumping.

Distinctions between Levels II and III

Children and youth in Level II are capable of walking without a hand-held mobility device

e after age 4 (although they may choose to use one at times). Children and youth in Level III need a hand-held mobility device to walk indoors and use wheeled mobility outdoors and in the community.

Distinctions between Levels III and IV

Children and youth in Level III sit on their own or require at most limited external support to sit, are more independent in standing transfers, and walk with a hand-held mobility device.

Children and youth in Level IV function in sitting (usually supported) but self-mobility is limited. Children and

youth in Level IV are more likely to be transported in a manual wheelchair or use powered mobility.

Distinctions between Levels IV and V

Children and youth in Level V have severe limitations in head and trunk control and require extensive assisted technology and physical assistance. Self-mobility is achieved only if the child/youth can learn how to operate a powered wheelchair.

A clinical evaluation includes history taking and neurological examination. Associated impairments should be evaluated by reviewing the history of true recurrent seizures, cognitive assessment, visual acuity, and hearing evaluation.

Management:

The goal of management of cerebral palsy is not to cure or to achieve normalcy but to increase functionality, improve capabilities, and sustain health in terms of locomotion, cognitive development, social interaction, and independence. The best clinical outcomes result from early, intensive management. A modern team approach focuses on total patient development, not just on improvement of a single symptom. Treatment programs encompass physical and behavioral therapy, pharmacologic and surgical treatments, mechanical aids, and management of associated medical conditions. In physical, occupational, speech, and behavioral therapies, the