Cerebral palsy

Objectives
1. Define cerebral palsy, tone, reflex
2. Discuss classification of cerebral palsy
3. Discuss etiology and prevalence of cerebral palsy
4. Describe the assessment and recording of examination of a patient with cerebral palsy
5. Discuss natural history of cerebral palsy
6. Discuss the effect of contracture of the gastrosoleus, hamstrings, and hip flexors
7. Describe methods of mobility and communication for children with spastic quadriplegia
8. Discuss current modalities used for control of tone
9. Discuss problems of parents of children with cerebral palsy

Discussion points
1. Is motion analysis necessary for optimum management of the cerebral palsy patient?
2. What do adults with cerebral palsy consider the most important factor in their assessment of treatment?

Discussion
Cerebral palsy is a very common childhood condition, with a consistent prevalence rate estimated at 2/1000 children in developed countries. There are several definitions, Bleck prefers that of Ingram "an inclusive term to describe a group of non-progressive disorders occurring in young children in which disease of the brain causes impairment of motor function...may be the result of paresis, involuntary movement or incoordination, ...motor disorders which are transient or the result of progressive disease of the brain or attributable to abnormalities of the spinal cord are excluded." A reflex is a simple motor action, stereotyped and repeatable, elicited by a sensory stimulus. Some motor responses are described as fixed action patterns. This is similar to a reflex, but is a more complex motor act, this is generated internally or elicited by sensory stimulus. An example of patterning is dorsiflexion of the ankle accompanying active knee flexion initiated while in the sitting position (confusion response). Tone is the state of partial contraction in which muscles maintain their posture without fully relaxing. It is muscle tone that permits one to lift an infant by placing the hands under the infant's arms; infants with low tone will collapse through the examiner's hands. A pathologic increase of tone is called spasticity; some writers use spasticity and tone interchangeably, but spasticity is the better term if pathologically increased tone is being described.
Cerebral palsy is usually classified anatomically as monoplegia, diplegia, (triplegia - rare, hemiplegia, or quadriplegia. Sometimes "paresis" is substituted for the "plegia" portion of the term, without any change in meaning. Children with diplegia have much greater involvement of the lower than upper limbs, but there are still some fine motor deficiencies in the upper limbs. Sometimes total body involvement is used interchangeably with quadriplegia. Children with hemiplegia usually have a focal lesion in one cerebral hemisphere, instead of a more global etiology. A physiologic classification designates the disorder as spastic, athetoid, ataxic, mixed, or hypotonic. Both classifications are usually designated in descriptions, for example, spastic diplegia.

Some causes of cerebral palsy are evident, such as very low birthweight with immaturity of the central nervous system, perinatal hypoxia or anoxia, or postnatal causes such as meningitis, vascular insults to the brain, child abuse, or near drowning. In other children, the etiology may be less obvious, such as exposure to toxic substances or as yet undelineated genetic factors. Although the musculoskeletal features of cerebral palsy are most evident, it is necessary to remember that visual, swallowing, gastrointestinal and genitourinary dysfunction, learning disorders, hearing problems, and emotional liability are all frequent in children with cerebral palsy.

Assessment of the child with cerebral palsy can be challenging. Quantifying range of motion can be especially difficult; examination of the same child will be quite different dependent on whether he/she is calm or upset. Perry has documented the imprecision of examination of spasticity. The end point is often indistinct; for example, when assessing dorsiflexion of the ankle, there may be an initial resistance, followed by a giving way of the gastrosoleus relaxing to another fixed endpoint. This is presumably a manifestation of the "clasp-knife" reflex, but which is the more significant measurement? The initial resistance is more likely what affects function but the presence of further movement eliminates contracture. We often talk of contracture of a joint or a muscle tendon unit when excursion is limited; without there necessarily being an anatomic contracture. For example, the Ely test for quadriceps contracture is performed by flexing the knee of the prone child, and observing the pelvis rock off the examining table. The same maneuver performed on the same child while asleep may show no apparent contracture. Of course, testing ankle dorsiflexion also is often dependent on whether the knee is flexed or extended. As spasticity persists, there will be secondary structural effects. This is most evident at the hip. The predominance of spasticity is always in the flexor muscle mass. As a result of the lower limbs external rotation of the limb bud, the flexor muscles are anterior at the hip, but posterior at the knee and ankle. At the hip, the longterm effects of untreated persistent spasticity of adductor and flexor tone is altered growth of the proximal femur into valgus (little traction on the greater trochanter from weak abductors) and anteversion, followed by subluxation, secondary acetabular dysplasia from the laterally displaced femoral head, and subsequent dislocation. This sequence of events is obviously more accelerated in the presence of more severe spasticity. This is an example of the effect of continuing structural changes from non-progressive cerebral dysfunction. In the upper limb, the flexors are anterior. Sensory deficiencies, including stereognosis have a much greater role in considering management of the upper limb, as the sensory component of the disorder is not affected by intervention.

The diplegic child is perhaps the prototype for orthopaedic evaluation an intervention. The diplegic child is ambulatory, with a crouch gait secondary to hip and knee flexion deformity, and equinus posturing of the ankle. Since an entire volume was devoted to the diplegic child, only the most basic of considerations follow here. Assessment must include gait, with attention to the
sagittal and coronal planes, as well as rotation. Typically, there is internal rotation of the lower limbs in addition to hip and knee flexion and ankle equinus. The goal for intervention is obviously elimination of contracture and rotational deviations. Hip flexor and adductor release and transfer are standard early measures, along with hamstring lengthening. Heelcord lengthening is a very common measure, but the penalty to the child following an over-lengthened heelcord is severe as he/she is unable to control forward thrust of the tibia during stance phase of gait, which will increase the crouch. Osteotomy, especially proximal femoral, is often performed later to correct subluxation and/or increased internal rotation. Orthotic control of the ankle is useful when there is no contracture or spasticity that precludes proper fitting. Motion analysis is very useful in assessing and quantitating angular and rotational deviations during gait. Dynamic EMG studies can implicate spasticity of certain muscles, such as the posterior tibial, as being primary in causing dynamic inversion and internal rotation of the foot during gait. Motion analysis is the only objective way of recording function prior to intervention and is becoming a prerequisite to meaningful documentation of the effects of intervention, both surgical and orthotic, in the diplegic child.

The natural history of the cerebral palseied child is obviously dependent of the type and severity of the motion disorder. We are still incapable of affording much meaningful intervention for athetoid movement. In addition to the surgical management of contracture secondary to spasticity, there are now systemic and regional approaches. Baclofen, either orally or intrathecally, is effective in reducing spasticity, with many fewer disabling side effects than previously used antispasmodic drugs. Rhizotomy has a limited but definite role for selectively reducing spasticity in the diplegic child; there are obvious limitations for the quadriplegic child. The quadriplegic child rarely can achieve meaningful ambulation, and mobility is thus dependent on wheelchair mobility. Mobility, activities of daily living, and communication, not ambulation, are the most desired goals of adults with total body involved cerebral palsy. Use of wheelchair hand and head controls, communication boards, and other computer assisted technology have expanded the horizons of the child and adult with total body involvement. Surgery on severely disabled patients with cerebral palsy is associated with a high complication rate.

Parents of children with chronic disabling conditions have unique problems which may not be readily understood by the professional. Mercer Rang has beautifully analyzed their problems and how the professional can meaningfully interact with them in his 1982 Easter Seal guide. To further distill his already succinct thoughts: The problem never goes away, crisis times are present throughout life, from the child's birth into the parents retirement years in that some type of assisted living arrangements must be made if the parent is no longer capable of caring for the child.

References


