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Surgery of the Upper Extremity in Children with Hemiplegic Cerebral Palsy

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Children with cerebral palsy and hemiplegia are special members of our family. While they have so much to offer, they often cannot reach their potential because of physical impairments. Limitation of hand function is only one of their problems, but an important one when the use of the hand for work and play and self care is considered. Doing an operation on a hand does not make a normal hand, but with the right indications, done well, and with good rehabilitation surgery can be of significant benefit.

A glossary of the medical terms used here is present at the end.

I - THE CONDITION OF THE HAND - While there are some exceptions, the usual effect that the brain injury of CP has upon the hand results in a typical positioning and limitation of the hand. The hand is usually held with: the elbow slightly flexed, the forearm held with the palm turned down (pronated), the wrist slightly flexed, and the thumb against the palm (adducted). (FIGURE 1). The muscles holding the segments of the arm in these positions are under the influence of an "upper motor neuron" type injury causing increased tightness (tone) that is not under the voluntary control of the person. When the person attempts to use the hand, the tone increases and the tightness of the positioning is accentuated (myotonic contracture). When the person sleeps, the tone often decreases. As the child grows bone length increases, but, lacking the usual stretching of normal, the muscles tightness become worse and can become fixed (myostatic contracture). While increased muscle tone in certain, specific muscles is the usual cause for problems in the CP child's hand, there may also be weakness of muscles that normally counteract the tight (spastic) ones.

II - TREATMENT - The ways we have to help with these problems are: exercises, splinting, medications, and surgery. I will deal with the first three briefly, since the purpose of this presentation is to give information about the surgery. But the first three, especially exercises and splinting, are essential parts of the treatment with or without surgery.

A. EXERCISES are passive stretching of tight muscles and strengthening of their antagonists (muscles doing the opposite of what the spastic ones do).

A1. PASSIVE STRETCH - Under normal conditions, all muscles are contracted and relaxed constantly during every day. Reaching for a glass on a shelf first stretches the elbow flexors, pronator, wrist flexors and thumb adductors at the same time contracting their opposites; grasping the glass strengthens thumb and finger flexors, and elbow flexors. This might be done dozens or more times a day and results in normal muscle lengthening and strength. The ideal goal of passive exercises (while impossible) is to reproduce this type of daily activity. When the child is young, the parent(s) and other family members can help by straightening the elbow, turning the palm up, and completely straightening the wrist and fingers of the child. In addition, The thumb can be pulled away from the palm. When the child is older, they can do the exercises themselves by crawling with palm on the floor, by doing "push-ups" against the wall, or by sitting on the hand and straightening the elbow. While constant exercise is impossible, doing these exercises once or twice a day for two or three minutes can make a tremendous difference.

A2. STRENGTHENING - Strengthening of antagonist of the spastic muscles can be useful to increase awareness of the involved hand and better positioning/use. But since the spastic muscles cannot be relaxed, it is impossible for this to fix the problem. The specific muscles that can and should be strengthened are the supinators (turning the palm up), and the wrist extensors (wrist curls with the palm down), and to a lesser degree, the finger extensors and thumb abductors (muscles pulling the thumb away from the palm done by pretending the fingers and thumb are the legs of a spider doing push-ups). Strengthening exercises are especially important after surgery has changed the effect of the spastic muscles.

B. SPLINTING – There are different approaches to splinting. Use of splints in the daytime can help position the wrist and thumb, but often large splints actually block the use of the hand and decrease the child's inclination to use the hand. Using a splint holding the wrist and fingers near full extension at night (while sleeping) can help stretch spastic muscles while bone growth is proceeding during 1/3 of the day. Use of a night splint is especially useful after surgery to protect muscles that have been moved to a new position to do a new function.

C. MEDICATION - Certain medications, taken by mouth or through surgically implanted infusion pumps, reduce muscle tone; and injection of Botox directly into a muscle will paralyze or weaken that muscle for 3 to 4 months. These treatments can help relax a muscle to make stretching and strengthening exercises easier, and Botox injection can be useful in determining the possible usefulness of surgery.

D. SURGERY

D1. INDICATIONS – The decision that surgery might help a specific child should be made carefully and after several observations. The injury in the child with CP is in the brain and, in addition to interfering with muscle control, can alter the sensibility of the hand and can make the child un-aware s/he even has that hand. Surgery of the arm/hand will not change this and is contra-indicated when the child ignores the hand, making no attempt at spontaneous use, even as a poor helper. Poor ability to feel with the hand (sensation) can be another reason that surgery is not indicated (one easy test is to place a dime or a quarter in the hand and ask the child which he is holding. If s/he cannot tell the difference, the benefit of surgery may be limited). If the child shows attempts at use of the hand, and is unable to use the hand because of muscle imbalance, operative treatment may be useful. The opinion of an experienced surgeon or therapist is needed. My simple evaluation involves giving the child a small container with a screw top and asking him/her to get a toy out of the container. If the child holds the container between his knees to get the top off, he probably will have limited benefit from surgery. If he uses his involved hand, even poorly, he is more likely to benefit from surgery. This awareness of the involved hand changes greatly as the child matures.

In addition to muscle spasticity, children with CP may have movement disorders called dystonias (prior names were athetosis, chorea, hemiballism). These movement disorders generally are

contraindications to surgery except when correcting associated spasticity is the reason for the surgery or when a joint fusion (arthrodesis) is the surgery proposed.

Another general rule regarding who might benefit from surgery is that children less than 4 or 5 years old are not surgical candidates. This is because the brain is developing and changing rapidly during the early years and the condition of the hand might change dramatically once the brain starts to mature. Also these children do not cooperate well with the rehabilitation after the surgery.

Some children will not cooperate with the surgery rehabilitation because of a lack of understanding or for emotional reasons. Some families cannot provide the support needed for surgery to be beneficial. These considerations are especially hard to assess, but are essential and misjudging them is the cause of many "failed" operations.

I feel that the age for surgery is when the examiner can be confident that a correctable problem is consistently present, when the child is old enough to make use of the surgery, and when the child is young enough to have the potential to incorporate the benefits of the surgery into his use of the hand. This is usually around the ages of 6 to 8 years old, but can be as young as 4 and can extend into adulthood.

Unfortunately we do not have reliable numbers about the success of surgery. Most of the time the hand is at least better than before the surgery. It is rare that the hand is made worse. The biggest problem is recurrence of wrist flexion positioning when wrist surgery is done on the young child since that surgery requires a great deal of early, complex therapy and requires that exercises and observation be continued until growth is complete. It is hard to keep up this prolonged effort. The other operations listed below almost never need to be "re-done". As the child grows, however, things change and additional surgery may be needed. Each surgeon differs on how many operations should be done at one time. Some think only one or two should be done to see the effect of those operations; others think everything should be done as once to facilitate the rehabilitation at one time.

D2. PRACTICAL MATTERS REGARDING SURGERY – hand surgery in CP children is usually done as an outpatient except for wrist fusion surgery which involves an overnight stay for pain relief. Most of the other operations seem to cause remarkably little pain. The surgery is done to improve function (not just appearance) and is covered by insurance.

D3. TYPES OF SURGERY – can be divided into: 1) weakens spastic muscles, 2) transfers a muscle to a new position and function, 3) fuses a joint.

WEAKENING A SPASTIC MUSCLE – muscles can be weakened by making them longer (loosening them), or by destroying part of their nerve supply.

The most common way to weaken a muscle is by lengthening or completely detaching its tendon or muscle from its attachment to bone. Cutting the tendon is a zig-zag fashion and sewing it in that position is intended to provide a specific amount of weakening so as to not completely lose its function. This is called a Z-plasty lengthening or Z-lengthening.

Neurectomy, or the cutting or crushing of specific motor nerve fibers, selectively destroys the nerve supply to a muscle and reduces the amount of muscle that is functioning, thereby weakening it. The actual effect of this surgery is hard to predict and this operation is not done in the United States very commonly.

TENDON TRANSFERS - The attachment of a tendon can be moved to a different place to do a better or different function. The most common example of this surgery is moving a tendon at the wrist to change it from a wrist flexor to a wrist extensor.

JOINT FUSION (ARTHRODESIS) – is done by removing the cartilage covers that make up a normal sliding joint and placing raw bone together to get them to heal in a fixed, stable and strong position. Common fusions that are done are of the wrist and middle joint of the thumb.

This operation is usually done only after the child is fully grown since the growth plate (potential) can be damaged during the operation. In older children with poor wrist and thumb control, or in children with dystonia, this is a very useful operation.

D4. SURGERY LISTED BY SPECIFIC AREA AND PROBLEM -

ELBOW – tightness of the elbow flexors can prevent good positioning of the hand. Surgery to weaken the elbow flexors is done by cutting the origins of the brachioradialis and brachialis muscles can be performed. The biceps is usually left intact since it is a forearm supinator, a function that best retained.

FOREARM – the pronation positioning of the forearm (rotation with the palm down and difficulty rotating the forearm so the palms of the hands face each other) can interfere greatly with two handed activities. The pronator teres muscle in the mid forearm can be cut, lengthened, or transferred. I think that transfer of the pronator so it circles the radius in the opposite direction and becomes a supinating force is particularly useful.

WRIST – flexion positioning due to spastic wrist and finger flexors interferes with being able to open the hand and spread the thumb from the palm. Several tendon transfers can be used here, along with lengthening of the wrist flexors. The most common are moving the wrist flexor (a spastic, deforming muscle) on the ulnar (little finger) side of the forearm to the dorsum (back) of the wrist to strengthen wrist extension. Another is moving the ulnar wrist extensor to the middle of the wrist giving it more mechanical advantage to extend the wrist. Once bone growth is complete, serious consideration is given to doing a wrist fusion if the wrist is not in a functional position. This provides a strong, stable foundation for finger and thumb function. In quadriplegic or double hemiplegic patients a wrist fusion can limit toilet care if the child is independent in this function because of the hand under consideration.

FINGERS – can be affected by tight long flexors that keep the fingers (especially middle and ring) curled, or by tight hand intrinsic muscles that keep the base of the fingers bent while causing the middle of the finger to bend backwards (swan-neck deformity). When severe, these either of these muscles can be cut, but some hand grip weakness is expected.

THUMB – the thumb is often held against or in the palm of the hand. Loosening the muscles (adductors and flexors) that pull the thumb there along with lengthening the skin of the thumb web space is helpful. This usually has to be aided by a tendon transfer to strengthen pulling the thumb away from the palm (abduction). Sometimes the middle joint of the thumb is loose and bends backward. In the young child, the joint can be tightened; in the older child the joint can be fused.

D5. CARE AFTER THE SURGERY – this is as important as the surgery itself. Tendon lengthening and transfers need about 5 weeks in a cast to heal enough to start rehabilitation. After the 5 weeks, the joints need to be moved, the new muscle functions need to be learned and strengthened, and use in everyday life needs to be learned. Initially the patient wears a protective splint full time except for therapy sessions to keep the hand in the best position. Once basic control has been achieved (usually 4-6 weeks), the hand is given some free time out of the splint. Strengthening exercises gradually make the hand more and more independent and the use of daytime protective splinting is reduced. However, a night splint is commonly worn for at least a year or often until growth is completed. During this time the child is best served by seeing a therapist to learn and practice the exercises at first 2 to 3 times a week. As the child recovers and home exercises are implemented, the therapy visits are reduced so that by the third month they are required only once a week or every other week. The development of a home exercise program is essential. This does not need to involve long sessions, but needs to be consistent. Surgery is not a magic cure, and if the family cannot honestly devote the daily attention that is needed, then it might be better to not do the surgery.

E. GLOSSARY

Arthrodesis: Joining together of the bones that normally comprise a joint. Done by removing the normal cartilage cover from the bones and holding raw bone ends together with pins, plate or cast until they heal as a broken bone would

Dystonia: Uncontrolled, spontaneous movements. More common example is the tremor of Parkinson's disease in adults.

Lower motor neuron lesion: paralysis of muscle produced by losing the end nerve fibers giving control of the muscle. Best examples are severing the nerve or polio

Upper motor neuron lesion: paralysis (or decreased control/strength) of muscle caused damage to nerve connections in the spinal cord or brain which preserves basic reflexes giving the muscle increased tone and increased sensitivity but not under voluntary control

Joint fusion: see "Arthrodesis"

Muscle/Tendon "release": Relieving a muscle of its tension or tone by cutting or lengthening or loosening the unit. Typically the upper end is cut free from bone or the lower end is cut in a zig-zag fashion and sewed in a longer position

Tendon transfer: (previously call a tendon transplant) moving the lower attachment of a muscle/tendon to a new location changing its pull or function.

Supination: rotation of the forearm (between elbow and wrist) to turn the palm of the hand upward, as in forming the palm into a vessel that could hold "soup"

Pronation: rotation of the forearm to turn the palm downward. Opposite of supination

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