The Upper Extremity in Cerebral Palsy: An Overview

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Abstract

Upper extremity functional impairment is a common problem in individuals with cerebral palsy. The level of the disability is very variable, with many individuals having functional use of the limb but with decreased dexterity. The Gross Motor Function Classification System (GMFCS) defines gross motor function as it primarily relates to the trunk and lower extremities. This does also relate to the upper extremity since the ability to do functional ambulation with an assistive device is also impacted by upper extremity function. Manual Ability Classification System (MACS) is a functional assessment similar to the GMFCS for mobility but focused on upper extremity manual use. The MACS evaluates how the upper extremity hand functions for activities of daily living as well as for more sophisticated activities. It considers both hands as a unit; therefore, if one hand is very highly functional, the score may be very high even though one limb has virtually no function. The International Classification of Functioning, Disability and Health (ICF) defines the causes of a disability that would prevent a specific activity from being accomplished. Therefore, based on the ICF guidelines, a child may be able to feed themselves because of the physical impairment caused at

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the level of body function and structure by not having an upper extremity or hand that is functioning at the level required for this activity. Furthermore, there may be individual personal factors such as lack of motivation as the cause of not self-feeding. The upper extremity disability in children with CP may have a significant impact on the individual's participation and ability to complete activities. The goal of this chapter will be to provide an overview of the disability including expected natural history and a review of treatment options.

Keywords
Cerebral palsy · Upper extremity · MACS · SHUEE · Surgery · Botulinum

Introduction

Upper extremity functional impairment is a common problem in individuals with cerebral palsy. The level of the disability is very variable, with many individuals having functional use of the limb but with decreased dexterity. The gross motor function classification system (GMFCS) defines gross motor function as it primarily relates to the trunk and lower extremities. This does also relate to the upper extremity since the ability to do functional ambulation with an assistive device is also is impacted by upper extremity function. The upper extremity impact on function is especially significant for GMFCS III level mobility as well as those children who are high functioning GMFCS IV. Manual Ability Classification System (MACS) is a functional assessment similar to the GMFCS for mobility but focused on upper extremity manual use. The MAC scale system evaluates how the upper extremity hand functions for activities of daily living as well as for more sophisticated activities. It considers both hands as a unit; therefore, if one hand is very highly functional, the score may be very high even though one limb has virtually no function. The upper extremity disability and children with cerebral palsy (CP) vary widely based on the individuals need in the context of their environment.

The International Classification of Functioning, Disability and Health (ICF) defines the causes of a disability that would prevent a specific activity from being accomplished. Therefore, based on the ICF guidelines, a child may be on able to feed themselves because of the physical impairment caused at the level of body function and structure by not having an upper extremity or hand that is functioning at the level required for this activity. There may also be environmental factors that preclude self-feeding such as the lack of available proper utensils or food with a texture or structure that the individual can manage. Furthermore, there may be individual personal factors such as lack of motivation as the cause of not self-feeding. The upper extremity disability in children with CP may have a significant impact on the individual's participation and ability to complete activities. The goal of this chapter will be to provide an overview of the disability including expected natural history and a review of treatment options.

Natural History

The GMFCS level of motor involvement also tends to provide some definition of upper extremity involvement. GMFCS I often have hemiplegic pattern CP. The hemiplegic limb may have a very wide variation of motor disability from being a limb that has a little functional ability to a limb with only mild clumsiness. The most common pattern of GMFCS II level functional children diplegic pattern CP. The upper extremity in children with diplegic pattern CP tends to show mild motor disability. Many children have only mild to moderate fine motor difficulties with diplegia. GMFCS level III usually have very functional upper limbs. There are a group of children with very asymmetric diplegic pattern involvement also called triplegic who may have one limb with a significant motor disability. This triplegic pattern involvement often makes functional use of a mobility aid difficult unless the motor function is such that the individual can walk with a single crutch or cane. For

GMFCS IV level of function, the often have most common difficulty is related to extremity motor problems limiting the use of an assistive device. Often, there is also a combination of trunk coordination problems as well as upper extremity function. GMFCS V level function is the individuals with the most severe upper extremity disability. It is very uncommon to have a GMFCS V level function with good upper extremity function, although there are children whose upper extremity provides very useful function for activities of daily living such as self-feeding and operating a power wheelchair.

Normal Development of Function of Children's Upper Extremities

Upper extremity spastic deformities start out as a clinched fist position with the thumb in the palm under the flexed fingers. This is an especially common posture in the affected limb of the child with hemiplegia but may also be seen with bilateral CP. As children grow, the fingers open first, and as more maturity and development occur, the thumb relaxes out of the palm. Often, in children with hemiplegia, the fingers are out of the flexed position by 2–3 years of age, and over the next several years, the thumb slowly relaxes. By 6–9 years of age, the thumb may be at the level of maximum abduction, and wrist flexion is becoming the predominant position. There is also significant elbow flexion with forearm pronation from early childhood. As children move through middle childhood and into adolescence, the elbow flexion and pronation often slowly decrease but almost never resolves completely but may become insignificant (Riad et al. 2007). By late childhood and early adolescence, the upper extremity deformity has developed the position it will maintain throughout the remainder of individuals' lives, except some of the contractures such as the contracted finger and wrist flexors may slowly become more fixed and more severe. These progressive contractures seem to be more common in quadriplegia than hemiplegia. Throughout childhood, the evaluation of individual children has to focus on their current function, physical deformity in the context of their age, and cognitive abilities. There is a great lack of published data describing the natural development of upper extremity function in children with CP. There is a large interest in correlating the brain imaging (Rose et al. 2011; van der Aa et al. 2013) with later function, and there is a renewed interest in imaging as it relates to enforced use therapy (Cao et al. 2015; Sutcliffe et al. 2009). Although there are theories how this imaging will be able to direct therapy and long-term prognosis (Friel et al. 2014; Holmefur et al. 2013; Spittle et al. 2009), there is not any real live data with long-term follow-up through to maturity to confirm these assumptions.

Classifying Upper Extremity Function

The MACS classification system of upper extremity function in children with CP has become the best recognized for overall monitoring. The disadvantage of this system is that it considers both hands as one unit and does not really separate the individual limb function. MACS is useful however to define overall upper extremity function in children with CP. It has good validity and reliability (Eliasson et al. 2006; Jeevanantham et al. 2015) Table 1.

We have use an upper extremity functional rating system that is directed at individual limbs. The goal of this system is to define the function and problems related to the individual limb. This classification system has not been validated; however, we find it useful to assess a limb (Table 2).

Another popular classification of function was published by House (House et al. 1981) related to function of the thumb and how the whole hand is utilized. The advantage of this classification is that it does focus on the individual hand (Table 3).

Other measures of hand function include the Assisting Hand assessment (AHA) and Children's Hand-use Experience Questionnaire (CHEQ) are instruments used to assess hand functional use

Table 1 Manual ability classification system (MACS)

Level I	Handles objects easily and successfully
Level II	Handles most objects but with somewhat reduced quality and/or speed of achievement
Level III	Handles objects with difficulty, needs help to prepare and/or modify activities
Level IV	Handles a limited selection easily managed objects in adapted situations
Level V	Does not handle objects and has severely limited ability to perform even simple activity

More detailed instructions are also available for down load: http://www.macs.nu/files/MACS_English_2010.pd

Table 2 Upper extremity function

Functional type:	
A	Extremity is not functional
B	Can use hand as a paperweight, pressure assist, or posting device; is able to swipe a toy and turn a switch on and off
C	Hand has mass grasp but poor active control
D	Hand has active grasp and release and can place an object with some degree of accuracy
E	Hand has fine pinch useful for holding a pen or pencil, has key pinch with the thumb
F	Normal function can be used for buttoning and shoestring tying, thumb has fine tripod opposition
Within each type, also assess level of contractures:	
I.	No contractures
II.	Dynamic contractures
III.	Fixed contractures

over time. These instruments seem to be most useful in the research environment since they are very time intensive to administer (Ryll et al. 2016). Further information on these instruments are available in ▶ Chap. 109, "Upper Extremity Assessment and Outcome Evaluation in Cerebral Palsy". Another instrument which can be used for diagnostic purposes is the Shriners Hospital for Children Upper Extremity Evaluation (SHUEE). The SHUEE testing is helpful to discriminate specific body impairments at the level of the hand which impact functional use (Davids et al. 2006). In this way, it is helpful for planning surgical treatment, orthotic use, or specific therapy interventions (▶ Chap. 109, "Upper Extremity Assessment and Outcome Evaluation in Cerebral Palsy").

Table 3 House functional assessment

Class	Designation	Activity level
0	Does not use	Does not use
1	Poor passive assist	Uses as a stabilizing weight
2	Fair passive assist	Holds object placed into hand
3	Good passive assist	Holds and stabilizes object for use by other hand
4	Poor active assist	Actively grasps object and holds weakly
5	Fair active assist	Actively grasps object and stabilizes well
6	Good active assist	Actively grasps object and manipulates against other hand
7	Spontaneous partial use	Carries out bimanual activities easily and occasional spontaneous use
8	Spontaneous use complete	Uses hand independently

As reported in Upper Extremity Chap. by Kozen, Chap. 34, p. 773 based on (House et al. 1981)

Treatment

Standard first-line treatment for upper extremity disability involves approaches using occupational therapy to encourage bimanual hand use in age-appropriate method (Charles and Gordon 2006). Constraint-induced movement therapy has become increasingly popular method to encourage the use of a unilateral CP upper limb. There are a wide variety of techniques including temporary splinting to complete long arm cast application of the well limb. Many short-term improvements have been reported (DeLuca et al. 2012); however, dosages and long-term impact are still unclear (▶ Chap. 179, "Constraint-Induced Movement Therapy for Children and Youth with Hemiplegic/Unilateral Cerebral Palsy").

The use of botulinum toxin has also been extensively reported in children with CP. The functional improvement with injection in the upper limb includes many reports that document short-term benefit (Sanger et al. 2007; Satila et al. 2006; Satila et al. 2006). There is no data on the long-term benefit from the use of botulinum toxin

in the upper extremity. There are other less commonly used methods to reduce muscle tone such as neurectomy and intrathecal baclofen (▶ Chap. 179, "Constraint-Induced Movement Therapy for Children and Youth with Hemiplegic/Unilateral Cerebral Palsy").

Surgical correction of the deformities has a long and well-established history. There are still however many variations in practice as it relates to surgical corrections of the upper extremity. When making surgical decisions and recommendations, it is very important to continue to consider the goals to be accomplished. It is important not to only focus at the impairment or body level dysfunction but one also needs to be very aware of the patient-specific goals for use of the limb. In the ICF model, we have to consider the personal factors. These personal factors which are very important are the goals of the patient specifically what the activity of functional use they would like to accomplish, are there cosmetic concerns as it relates to the limb, and most importantly does the patient have an interest in changing his current limb position or function. If the child is very young and these decisions are made by proxy of the parent or caregiver. An issue in considering upper extremity surgery should be whether the patient themselves can have input. This raises the question of when is the best age to consider operative surgical reconstruction. One train of thought is that surgery should be done young in 4 or 5-year-old children so they get the benefit of the reconstructed limb to maximize its function. Surgery in the young child risks a higher rate of recurrent deformity. Another approach is to wait until the individual is an adolescent and then involve the individual in the decision-making as to what they would like to have accomplished. Surgery at this age also appears to have less risk of recurrent deformity and therefore better long-term maintenance of correction. There are no studies currently that give guidance on the timing related to age, although I favor waiting until the individual child can assist in the decision-making.

Specific Treatments

The current standard of upper extremity surgery in children with CP is to make an evaluation of the whole limb and then combine all the surgical treatments at one setting. This is the single- event multilevel surgery (SEMLS) approach that has become the standard of care for the lower extremity and is now also the standard of care for the upper extremity (Smitherman et al. 2011) (▶ Chap. 112, "Single-Event Multilevel Surgery for the Upper Extremity in Cerebral Palsy").

Specific areas which one should consider for surgical correction include the shoulder and elbow. The most common shoulder problems are contractures that limit abduction and make activities of daily care difficult in patients mostly with GMFCS V level who require attendant care. Improvement in ease of care is greatly facilitated by the release of pectoralis contractures (Domzalski et al. 2007). The primary issue at the elbow is a flexion contracture mainly creating cosmetic concerns except in some children with GMFCS V develop severe contractures which make custodial care and dressing difficult. Release of the biceps or brachialis usually improves this contracture. Children with severe pronation may develop radial head dislocations which seldom become painful and the only treatment should be consideration of radial head resection at skeletal maturity if they are symptomatic (Abu-Sneineh et al. 2003) (▶ Chap. 113, "Shoulder and Elbow Problems in Cerebral Palsy").

Pronation contractures are most common in the distal forearm. These pronation contractures sometimes are helpful in that they allow the hand to be in a position where he can operate a keyboard. These pronation contractures sometimes are helpful in that they allow the hand to be in a position where he can operate a keyboard. Care should be taken to avoid placing the hand and forearm in the supinated position because this is both cosmetically and functionally much less appealing. Wrist flexion deformities and

contractures are very common. The Green transfer (Green and Banks 1962) of the flexor carpi ulnaris to the extensor carpi radialis brevis continues to be a mainstay treatment for this very common deformity (Beach et al. 1991). For individuals who have a nonfunctional hand, the use of a wrist fusion to place the hand and an optimal cosmetic position also eases care especially for placing the limb in shirt sleeves. The wrist fusion should be avoided in individuals who are using wrist motion to facilitate finger grasp or finger release (Thabet et al. 2012).

Thumb adduction is another common disability in the hand of the individual with CP. The thumb adduction may limit the ability to grasp objects in the palm or to use pinch grasp effectively. Most individuals with CP are able to use key pinch but not tripod pinch grasp because of the limited abduction motion present in the thumb. Thumb positions have been classified by House (House et al. 1981) with Type 1 being adduction with normal MCP and IP joints and Type 2 as adduction with flexion of MCP and IP joints that are not fixed. Type 3 has hyperextension of the MCP joint and abduction of the thumb, and Type 4 has a fixed flexion contracture of the MCP joint with severe abduction. Treatment of the thumb deformity requires releasing fixed contractures, stabilizing hypermobile joints, and augmenting weak muscles (▶ Chap. 114, "Forearm, Thumb, and Finger Deformities in Cerebral Palsy").

Complications

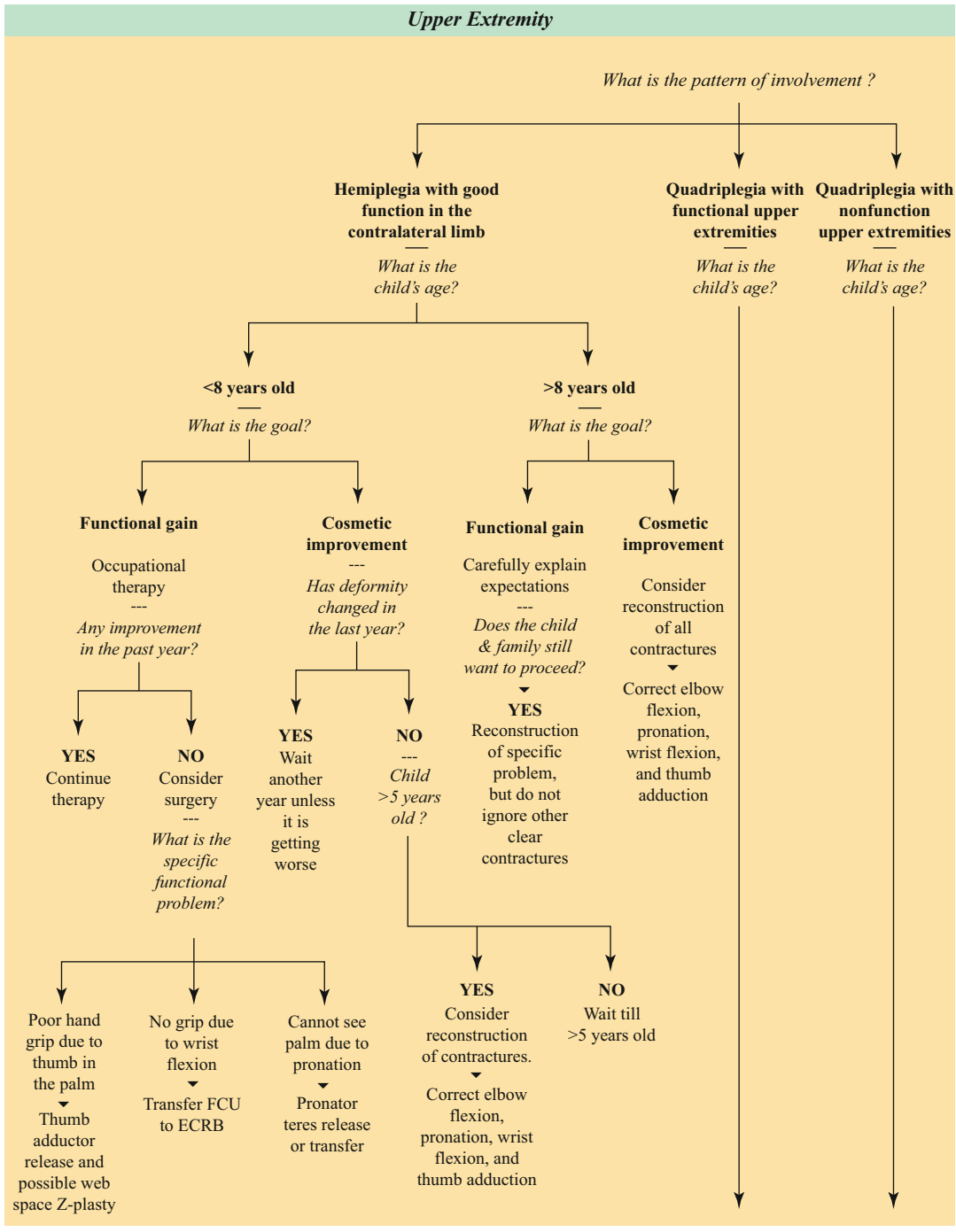
The main complication of treatment of the upper extremity in children with CP is that a lot of treatment may be given without much benefit. This is especially related to mild invasive treatments such as botulinum toxin, constraint therapy, and very intense therapy. Research needs to focus especially on the impact of these treatments long-term and also consider the negative impacts on the patient and family. The most major complications of surgical reconstruction are to not clearly defining the expected outcome goals to the patient and the family; often there is a feeling that expected goals therefore were not achieved. A common scenario is the adolescent very much wants surgery to improve the cosmetic appearance of his hemiplegic limb; however, the parents' goal is for the adolescent to start using the limb in a more functionally normal way. When this does not happen the parent is not satisfied but the adolescent is.

Clearly, another complication risk with upper extremity's surgery is recurrent deformity. This is especially likely in children who are very young when they have surgery. Another risk is overcorrection of a previous deformity. This is especially true for overcorrection of wrist flexion deformity which may then gradually become wrist extension deformity. Another, possible overcorrected deformity is forearm pronation especially when there is too much correction of the pronation, one can develop a supination contracture or deformity which makes families and patients very unhappy.

Conclusion

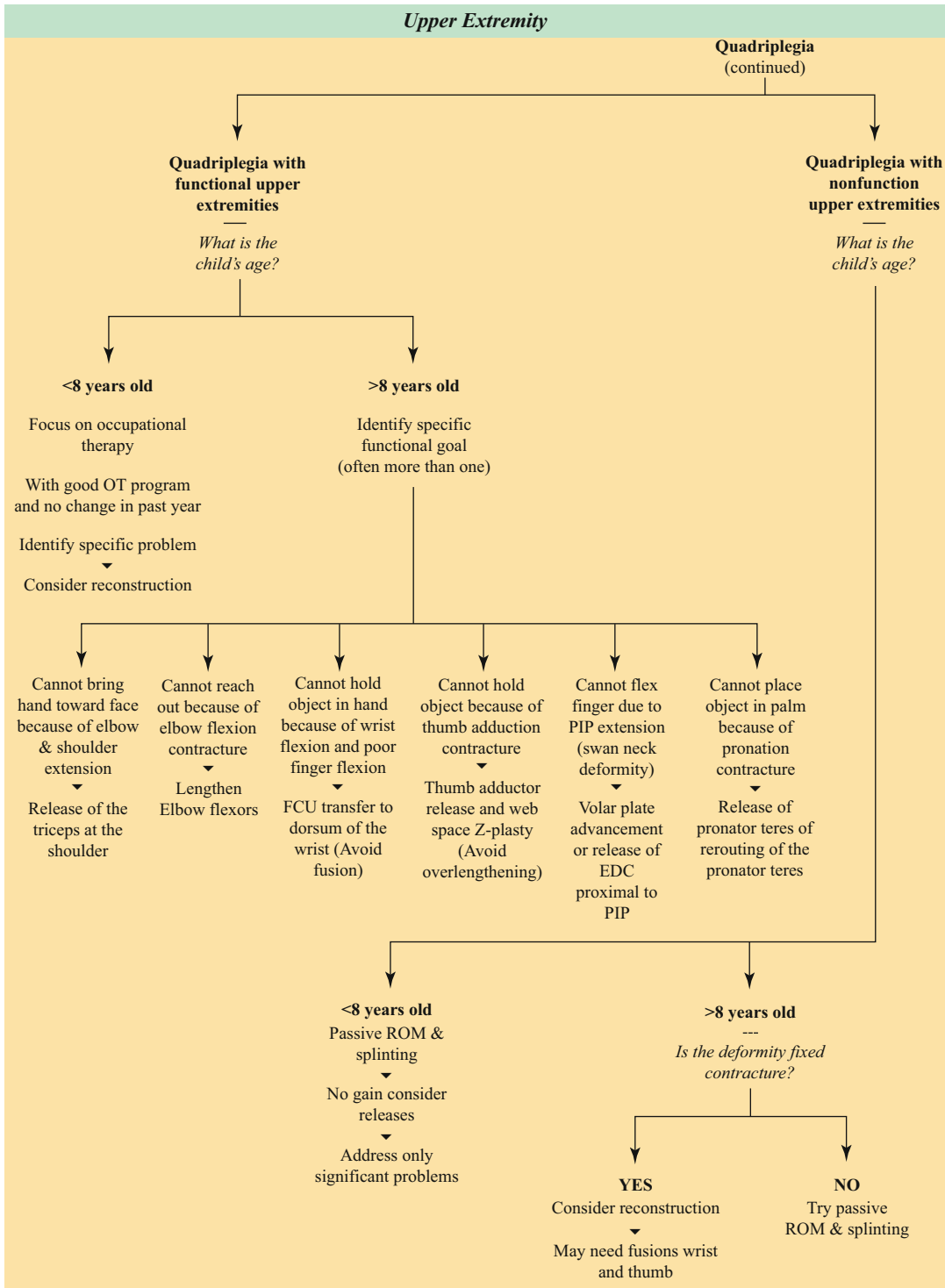
The classic child with a spastic upper extremity, in whom surgical treatment is considered, has spastic hemiplegia causing posturing of the involved upper extremity with the elbow flexed, forearm pronated, the wrist and fingers flexed, and the thumb adducted and flexed in the palm. Children with movement disorders (athetosis or dystonia) may present with upper extremity involvement; however, surgical correction is rarely indicated. The greatest task is to clearly define the functional difficulties (if any), determine optimum goals for a specific child's developmental stages, and bring together realistic long-term goals between patients, parents, and orthopedic surgeons. This task requires that surgeons understand the concerns of families and children especially the cosmetic concerns of the extremity and the specific functional concerns. Often, the concerns of patients, especially adolescents, are different from the concerns of parents. Also, orthopedists have to understand each component of the global

Table 4 Upper extremity algorithm



(continued)

Table 4 (continued)



extremity's impairment and how these impairments evolve with developmental maturation. It is important to have an algorithm conceptually when evaluating the child for surgery (Table 4).

Cross-References

- Constraint-Induced Movement Therapy for Children and Youth with Hemiplegic/Unilateral Cerebral Palsy
- Forearm, Thumb, and Finger Deformities in Cerebral Palsy
- Physical Examination and Kinematic Assessment of the Upper Extremity in Cerebral Palsy
- Shoulder and Elbow Problems in Cerebral Palsy
- Single-Event Multilevel Surgery for the Upper Extremity in Cerebral Palsy
- Spasticity, Dystonia, and Athetosis Management in the Upper Extremity in Cerebral Palsy
- Upper Extremity Assessment and Outcome Evaluation in Cerebral Palsy

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