Current Orthopaedics (2003) **17**, 105–116

doi:10.1054/cuor.2003.0329

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Current
ORTHOPAEDICS

MINI-SYMPOSIUM: CEREBRAL PALSY

(iii) Cerebral palsy of the upper extremity: the surgical perspective

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Summary This article describes the surgical management of cerebral palsy affecting the upper extremity. It starts with the clinical picture and its assessment using a team approach and prospective data collection. Established surgical treatment is described with emphasis on its mechanical basis. There is additional detail of the author's preferred techniques for surgery and early mobilisation for postoperative rehabilitation.

INTRODUCTION

Cerebral palsy (CP) is the most common childhood cause of upper extremity motor disability, with incidence rates of 1.5–3.0 per 1000 live births reported.[1] The incidence is higher in lower birthweight groups and appears to be rising over time. The Surveillance of Cerebral Palsy in Europe working group listed five key elements from 10 treatment centres across Europe for a diagnosis of cerebral palsy as: CP is a group of disorders (i.e. it is an umbrella term); it is permanent but not unchanging; it involves a disorder of movement and/or posture and of motor function; it is due to a non-progressive interference/lesion/abnormality; this interference/lesion/abnormality is in the developing/immature brain.[1]

CLINICAL FINDINGS

The European classification of cerebral palsy further subdivides CP into spastic, ataxic and dyskinetic types, which can coexist in an affected extremity.[1] Spastic and mixed types make up over 60% of the affected upper extremities.

Spastic CP

Spastic CP (also referred to as pyramidal CP) is characterised by increased muscle tone, increased reflexes and an abnormal pattern of posture and/or movement. The classic appearance is of internal rotation and adduction at the shoulder, flexion of the elbow and wrist, flexion of the fingers and a thumb-in-palm deformity (Figs. 1 and 2). The clinical picture is governed by a combination of increased tone in some muscle groups, weakness or complete paralysis in other groups and fixed or reducible joint positions secondary to these muscle imbalances. Fundamentally, there is a variable loss of control of muscle groups. Spasticity is typically abolished by sleep and general anaesthesia and exacerbated by rapid manual stretching during examination, by some splints, by emotional factors and by attempts to utilise the extremity or maintain a given position. Age has an important influence on the clinical picture. In very young children (under 2 years) CP may not be recognised. A tendency for the thumb to stay in the palm is one of the earliest signs. In older children weakness of wrist extension is apparent. Joints are frequently passively reducible to a functional position and abnormal grasp patterns are starting to develop. By late childhood and adolescence the classic posture of spastic CP is apparent with established contractures at the elbow and wrist and the presence of swan-neck and thumb-in-palm deformities. By adulthood fixed joint contractures are the rule and use of the limb may be restricted to a brachiothoracic grip.

There is a wide variation in severity however and some patients will present with few or milder deformities and reasonably good function. In spastic CP soft tissue procedures in the form of tendon transfers, musculotendinous unit lengthening and joint releases form the mainstay of surgical intervention.

Ataxic and dyskinetic CP

These latter two groups are also referred to as extrapyramidal CP and the hallmark of these types is the loss of

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control of muscle groups. Ataxic CP is characterised by loss of orderly muscle coordination. Dyskinetic CP may be dominated by reduced activity with increased tone (dystonic subtype) or increased activity coupled with reduced tone (choreo-athetotic subtype). Joint ranges are often preserved as the limb switches from one posture to another (Fig. 3).

Soft-tissue procedures are generally unhelpful in the purer forms of dyskinetic CP because of the lack of voluntary control. They may be helpful in mixed forms of CP if directed at the spastic component. The mainstay of surgical treatment in the dyskinetic group is joint fusion to introduce stability.

PREOPERATIVE ASSESSMENT

Team approach

The management of patients with CP requires the skills of many disciplines. The team includes the coordinating physician, the parents or carers of the patient, therapists and social workers as well as the surgical team. Following surgery, occupational and physiotherapists work with the patient and the patient's carers to maximise the benefits of the procedures. When emotional instability, behavioural problems or fear of intervention are present, the input of a clinical psychologist is invaluable.

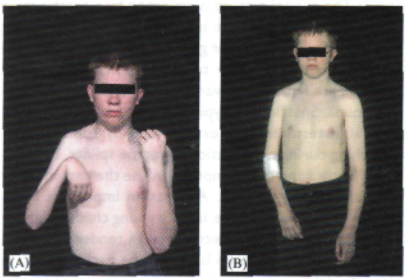
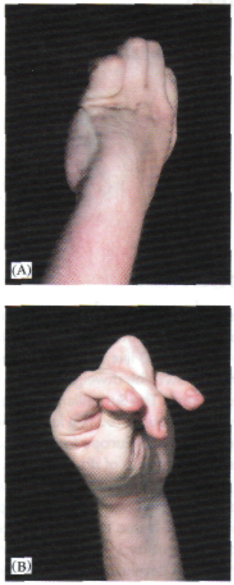


Figure 1 (A, B) Typical posture in CP, improvement after surgery.



Figures 3 (A, B) Changing postures in athetotic CP. Note the position of the finger metacarpophalageal joints.

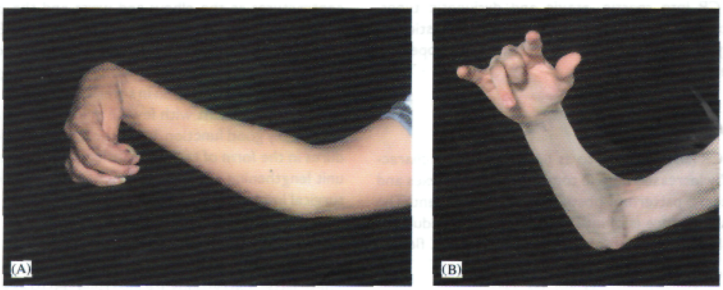


Figure 2 (A) Severe wrist flexion and thumb-in-palm deformities. (B) Elbow contracture.

General assessment

Delineating the goals of surgery to the upper extremity demands an assessment in the context of the patient's general condition. Surgery requires extra input from carers in terms of general support, transport to and from the hospital, participation with therapy, extra care with activities of daily living and increased psychosocial support during what can be a difficult period of time. It is crucial to identify and deal with any issues in these areas before embarking on surgery. Information from the carers provides an insight into how the upper extremity impacts on activities of daily living such as hygiene, balance, sitting and walking. The general neurological condition of the patient is assessed to formulate realistic goals on an individual basis, taking into account cognitive ability, hearing, speech and visual impairment against a background of the patient's age and environment. Mental impairment is no more a contraindication to surgery than young age as long as a patient is cooperative and motivated. It is important to appreciate that in some instances the goal of surgery may be directed towards the carers to enable them to look after the patient, for example, the treatment of severe contractures to facilitate hygiene, dressing and transport. Similarly the severity and the extent of CP are not necessarily contraindications; a patient with involvement of both legs may benefit greatly from even modest functional improvement in the upper extremity. Age becomes an important factor in that it may determine whether or not a patient will adapt to changes in an extremity. The cosmetic aspect of a deformity should not be underestimated as many adolescents and adults initially seek treatment for this purpose alone (Fig. 1).

Upper extremity assessment

A detailed baseline assessment of each affected upper extremity is carried out. As well as recording joint positions, range and power we use a combination of existing classifications to record limb control and the pattern of limb use (Table I). The reader is referred to these classifications for a detailed description of each category and grade. Photography and video are used to provide a clear record of preoperative posture and function.

Sensation

Sensory impairment is a common finding in CP, yet one of the largest patient series spanning 25 years and 718 operations did not find it a predictive factor for improvement after surgery.[6] Others have shown that stereognosis improved after surgery.[7] We record stereognosis using a pick up test and sensory thresholds using monofilaments (Table I) prospectively to investigate this further.

Table I. Upper extremity assessment in CP

Assessment	Areas assessed
Resting position	All joints
Active range	All joints, control, spasticity
Passive range	All joints
Hand placement[2]	Shoulder, elbow
Voluntary motor capacity[2]	Forearm rotation, wrist, fingers
Hand assessment[3]	Grasp, release, independence
Thumb deformity[4]	Thumb
Stereognosis[5]	Sensation
Semmes-Weinstein monofilaments	Sensation
Summary description of limb use	Whole limb
Photography and video	Whole limb

Electromyography

This investigation is difficult to perform accurately and reliably, particularly in a group of patients in whom involuntary contraction occurs so readily or in the young. Its use is based on the premises that muscles that fire out of phase or are non-synergic may not produce successful transfers, that these qualities are fixed and unaffected by surgery and that the ability to voluntarily contract muscle cannot be assessed by clinical examination. There is limited evidence to support these premises and having found little to be gained by electromyography over careful clinical examination, I have now abandoned it as an assessment tool.

Botulinum toxin A

Botulinum toxin injected into a muscle causes a reversible paralysis and in selected patients may be temporarily therapeutic.[8] Preoperatively it may help to establish which muscles are responsible for a given deformity and provide information about the expected effects of surgery. This however requires accurate, selective injection which is difficult to achieve, particularly in children. I no longer routinely use botulinum toxin for these reasons.

Splinting

Splints are cumbersome and may stimulate increased spasticity (often painful) and cover sensate skin. There is no logic behind night splinting unless it is part of a programme to correct a static joint contracture as spasticity is abolished by sleep. Most patients who present for surgery have tried and abandoned splints in the past. Daytime wrist and first web splinting prior to surgery both facilitates and demonstrates the effect of surgery. Postoperative splinting is discussed below.

SURGERY

The principles of surgery are as follows.

Define

Define the surgical aim at a given joint and identify the prime forces that give rise to the deformity. Note that these forces may be the result of an attempt at function. An example of this is the patient with a flexed wrist who has hyperextension deformities of the digits evident on attempts at grasp and release. Lengthening or release of the extensors is likely to worsen the deformity, whereas rebalancing the wrist often solves the problem and may demonstrate that the digital extensors are actually weak and require augmentation. It is vital to recognise the interrelationship between different joints when planning and staging surgery.

Restore

Restore the correct joint position, keeping in mind that some function in the deforming muscle or muscle group is essential if balance is to be maintained and an opposite deformity avoided. For this reason I prefer to perform lengthening procedures to releases. Where this is not possible, function can be preserved by releases at a musculotendinous level or at the muscle origin rather than at the tendon insertion.

Rebalance

Rebalance the joint by a transfer that will prevent recurrence of the original deformity. This requires adherence to the principles of tendon transfers whenever possible (Table 2) and a knowledge of the relevant muscles, their relative fibre lengths (a measure of excursion) and tension fractions (a measure of strength) (Table 3).

It is the application of these principles to the specific deformities encountered that constitutes the surgical plan and this is now described.

Table 2. The principles of tendon transfers

Correct joint contractures
Select a muscle of adequate strength
Select a muscle of adequate excursion
Select one tendon for one function
Select an expendable donor
Establish a straight line of pull
Position and time transfers so that they lie in tissue of optimal condition

Table 3. Forearm and hand relative values for measures of excursion and strength

Muscle	Mean resting fibre length (cm)	Tension fraction(%)
ECRL short (wrist) fibres	6.3	3.5
ECRB	6.1	4.2
EDC all fingers	5.8	5.5
ECU	4.5	4.5
FCR	5.2	4.1
FCU	4.2	6.7
FDS index	7.2	2.0
FDS middle	7.0	3.4
FDS ring	7.3	2.0
FDS little	7.0	0.9
PT average	5.1	5.5
EPL	5.7	1.3
EPB	4.3	0.8
APL	4.6	3.1
AP	3.6	3.0
FPB	3.6	1.3
PL	5.0	1.2
Brachioradialis	16.1	2.4

The shoulder

Internal rotation and adduction is frequently encountered at the shoulder but rarely warrants surgical intervention. In some patients the administration of botulinum toxin A into the pectoralis and subscapularis muscles may be beneficial both as a diagnostic and therapeutic option. Occasionally the deformity prevents adequate hand placement or hygiene and surgery is necessary. Restoration of glenohumeral range is achieved by lengthening of the subscapularis and pectoralis major muscles. A deltopectoral approach is necessary to access these when there is poor access to the axilla; otherwise, my own preference is an axillary approach. The subscapularis origin is released from the scapula and the pectoralis major tendon step-lengthened. The anterior shoulder capsule is preserved for stability. Rebalancing is achieved by transfer of the latissimus dorsi and/or teres major to the greater tuberosity accessed via a posterior arm crease incision.

The shoulder is splinted in external rotation and mid abduction for 4 weeks before commencing range of motion exercises.

In severe cases with posterior subluxation, an external rotation osteotomy has been advocated,[10] although this runs the risk of increasing the lever arm for internal rotation if unchecked by functioning external rotators.

Very occasionally a patient presents with an external rotation contracture and an abducted shoulder. Treatment options are deltoid release, lengthening of the external rotators and plication of the anterior capsule and subscapularis.[10]

The elbow

The deformity at the elbow is a flexion contracture due to spasm of the biceps, brachialis and brachioradialis. The first component of this is a fixed joint contracture which may be modest in young children but in older children and adults is typically around 30° from full extension. The second component is a dynamic contracture, which may be over 90° and which can be overcome passively to the level of the fixed contracture in the right conditions (Fig. 2B). This manifests itself as both an inability to extend the forearm into space and most significantly, to keep it there without rebound flexion. It is this latter problem that severely restricts function and lends itself to surgery with a predictably good result. In planning surgery on the elbow it is important to consider the impact that this will have on the wrist and the digits. Successful lengthening of the elbow flexors will lead to tightening of the flexors that take their origin from the medial epicondyle. For this reason I rebalance the wrist and fingers at the same operation but carry out the elbow surgery first.

Surgery consists of a lengthening of the elbow flexors (Figs. 4 and 6). Persistent contracture following this is due to the joint capsule and neurovascular axes. When the nerves and vessels are tight no further surgery can be expected to increase range. If the capsule alone is tight but the contracture no greater than 30°, I do not proceed further as experience has shown that useful function and control has been achieved without the counterproductive effects of severe postoperative pain from a joint release. When a severe fixed contracture persists the anterior elbow capsule is released from the humerus starting proximally, until a satisfactory range has been achieved.

The wrist and digital extension

In the wrist the deformity is of flexion with or without pronation and ulnar deviation (Fig. 2A). The deforming forces are complex and are mirrored by the variety of surgical options. The posture of the digits is intimately related to the dynamics of the wrist and so these are considered together. A systematic approach to analysis

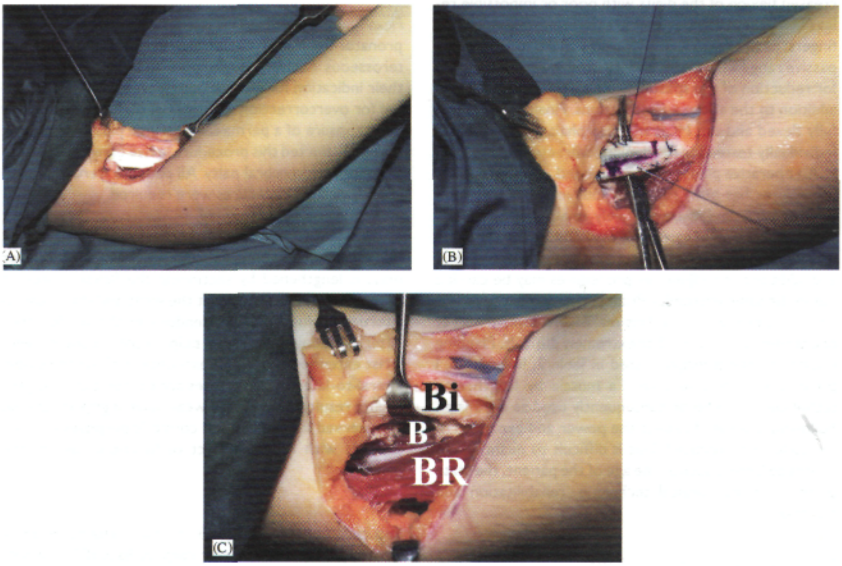


Figure 4 Elbow release. (A, B) The biceps tendon is exposed and step-lengthened via a transverse approach. (C) Musculoaponeurotic lengthening of the brachioradialis (BR) and brachialis (B) muscles is achieved by excision of a strip of the investing fascia and division of intramuscular tendinous fibres at this level (Bi = biceps).

and treatment is essential to avoid ineffectual treatment, worsening of the deformity or the creation of one that is different but equally incapacitating. The position of the wrist is key to hand function. In the flexed position the digital flexors have a poor mechanical advantage, tenodesis across the wrist may put the digits into an extended position which is exacerbated by attempts to reach and may lead to swan-necking (Fig. 7). Alternatively the fingers may be in a flexed posture making grasp impossible. In extreme or longstanding cases flexion, pronation and ulnar deviation at the wrist can give rise to a reverse (ulnar) grasp pattern.

A patient who consistently presents with the wrist in the flexed posture clearly has an imbalance at this joint with or without a fixed flexion contracture. The latter is assessed by testing passive extension at the wrist. If the wrist can be brought to neutral or beyond, wrist joint release is unlikely to be beneficial. Active wrist extension to neutral that cannot be maintained without a drift back into flexion or which collapses on attempts at digital flexion almost invariably indicates the need for wrist extensor augmentation.

With the wrist stabilised in neutral or beyond, digital extension is tested actively. Unless good active metacarpophalangeal (MCP) joint extension is obtained, digital extensor augmentation will be necessary to prevent increased flexion of the digits with poor or impossible release. In the absence of full active extension of the fingers, passive extension is tested. Failure to achieve full passive range with the wrist in neutral indicates the need for reduction of the spastic flexor forces on the digits in addition to the above augmentations. Finally, the wrist is fully flexed and the MCP and interphalangeal (IP) joints individually tested for fixed joint contractures.

An operative plan is now formulated. A wide range of procedures has been reported, but in any given study, the numbers tend to be relatively small, the criteria for surgery may vary, a variety of assessment and outcome scales are used, a variety of surgical techniques are used and additional or adjunctive procedures may be carried out in the same extremity. Prospective comparative studies are a distinct rarity. This makes the comparison of one technique with another extremely difficult. Planning is aided by the principles listed in Table 2. In addition, I prefer not to interfere with a muscle by lengthening techniques if it is to be subsequently used as a transfer. Following transfer I expect the power and excursion of a muscle to be reduced. This is difficult to quantify and relates to factors such as the age of the patient, the state of the muscles, surgical technique, rehabilitation and scarring.

Reduction of flexor deforming forces

Musculoaponeurotic lengthening. This is indicated for milder tightness without fixed joint contractures. There is little risk of over-lengthening and the technique is simple. A 2 cm wide strip of the antebrachial fascia is excised over the flexor-pronator mass, followed by division of the intramuscular fibrous septa, except in those muscles destined for transfer.

Musculotendinous lengthening. This procedure is carried out 2 cm proximal to the musculotendinous junction to lengthen without defunctioning. The tendon is exposed by gently spreading the muscle fibres and divided sharply. Steady gentle extension force typically gains 1–2 cm of lengthening.

Tendon step-lengthening. This procedure is indicated when severe tightness persists, for example when passive thumb IP joint extension is limited. A step-cut is made in the exposed tendon. The length of the step-cut is theoretically half of the increase in tendon length. Gerwin[11] recommends 0.05 cm lengthening per degree of joint contracture correction and tensioning of the wrist to the neutral position. Repair by tendon weave is both strong and allows tensioning but reduces the gain in length. My preference is a multistrand end-to-end repair (Fig. 4).

Flexor-pronator slide. This is a detachment of the flexor-pronator mass from the medial epicondyle, ulna and interosseous membrane. Zancolli and Zancolli[12] defined their indication for this major release noting the potential for overcorrection with loss of digital flexor power and exposure of a permanent intrinsic plus contracture. Pinzur[2] reported this procedure in five carefully selected patients with cerebral palsy. All patients were improved but adjunctive procedures were required.

The superficialis to profundus transfer. The digital flexor mass is lengthened by sectioning the flexor digitorum superficialis (FDS) tendons at the wrist and the flexor digitorum profundus (FDP) tendons in the midforearm. After tensioning to allow pulp-to-palm contact with wrist extension and digital extension with wrist flexion, lateral-to-lateral tenorraphies are carried out. This procedure results in significant weakening of grip and should be reserved for hygiene and cosmetic purposes in those patients with little prospect of functional use of the hand.

Reduction of flexor strength by release. The flexor carpi ulnaris (FCU) or the flexor carpi radialis (FCR) should not be released together as an extension deformity may follow at the wrist. I prefer to restrict either release to its use as a transfer.

Wrist extension augmentation

An understanding of the normal role of the tendons that cross the wrist is essential in evaluating the available transfers and releases. Dedicated wrist motors, FCU, FCR, extensor carpi radialis longus (ECRL), extensor carpi radialis brevis (ECRB) and extensor carpi ulnaris (ECU) are distinguished from the muscles whose tendons cross the wrist on the way to the digits. Whilst the latter also stabilise and move the wrist, they cannot be relied upon to do this when acting primarily on the digits.

The ECRB is chosen in preference to the ECRL as the recipient tendon because the ECRL extends and radially deviates the wrist and is an elbow flexor. In contrast the ECRB is a more effective wrist extensor because its moment arm for wrist extension is larger and that for radial deviation smaller, it is stronger than ECRL and the whole muscle is available, as it has no effect on the elbow.

FCU to ECRB transfer. This popular transfer simultaneously reduces the flexor and ulnar deviating force at the wrist and transfers this to the extensor side and has been reported to improve function.[6, 7, 13] The transfer can be routed directly through the interosseous membrane, around the ulnar side of the forearm in an attempt to increase supination or around the radial side in an attempt to reduce ulnar deviation.

The strength of the FCU is more than adequate in theory, though my own experience is that this muscle is frequently scarred and fibrotic in older children and adults. The mean fibre length is only two-thirds that of the ECRB and this may be reflected in the limited arc of flexion–extension that has been reported.[7, 13] Furthermore, release of the FCU removes a strong flexor of the wrist and if there is weakness or poor control of the FCR, wrist flexion can be severely weakened or lost. The FCU is a prime ulnar wrist stabiliser and Brand describes how its loss may be significant.[14]

ECU to ECRB transfer. Of the tendons that cross the wrist the ECU is unique in that its tendon has a constant relationship with the ulnar head.[9] Brand argues that forearm rotation into pronation changes the relationship of ECU to the flexion–extension axis resulting in a loss of the moment arm for wrist extension. This and a barely adequate excursion largely limit its use to situations in which wrist extension is present to neutral position with a tendency to ulnar deviation. It should be avoided when there is inadequate wrist extension. Procedures that

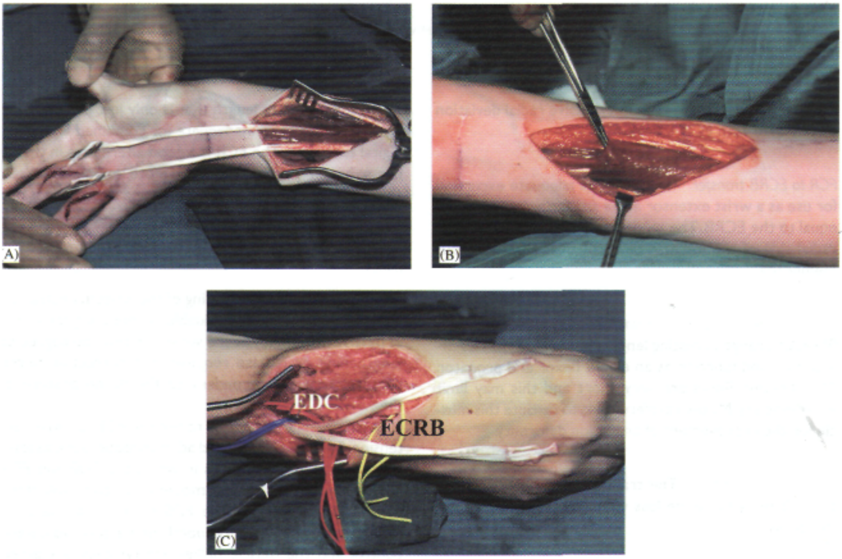


Figure 5 Interosseous transfer of the FDS for wrist and digital extension. (A) Harvested FDS tendons (the finger approach for simultaneous FDS tenodesis is shown). (B) Tendons passed through the interosseous membrane. (C) Tendons in the extensor compartment in preparation for weave to the ECRB and EDC proximal to the extensor retinaculum.

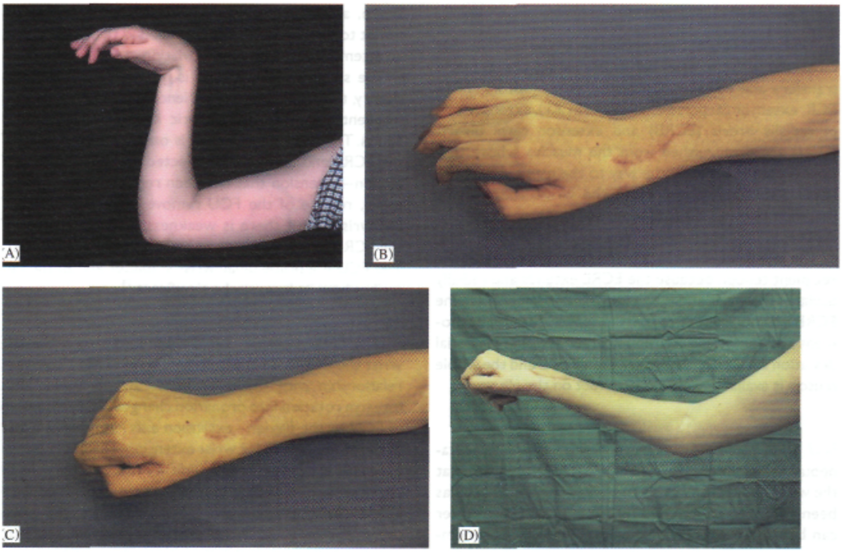


Figure 6 (A, B, C, D) Preoperative posture and early postoperative results of the patient shown in Figs. 4 and 5, digital extension and flexion with preservation of wrist extension, elbow extension.

weaken the FCU may result in excessive radial deviation if combined with ECU transfer.

FCR to ECRB transfer. The FCR has adequate excursion for use as a wrist extensor and a strength that is almost equal to the ECRB. The tendon can be taken around the radial side of the forearm or through the interosseous membrane.

Pronator teres (PT) to ECRB transfer. This muscle contains fibres of a range of resting lengths in-keeping with its orientation and function as an elbow flexor and pronator. The shortest fibres are very short and this may limit wrist mobility. My own preference is to reroute this muscle for the improvement of pronation deformity.

FDS to ECRB transfer. This transfer has ample excursion even allowing for some loss when transferred but may lack power.

Digital extension augmentation

The FCU, FCR and FDS have been used as transfers to augment digital extension. The FCU has adequate strength but a fibre length that is only just adequate for wrist extension and inadequate for combined wrist and digital extension. The FCR is a good alternative in that it has a better excursion and is more expendable (see above). The FDS transfer has ample excursion and is described below.

Authors preferred method of treatment of the wrist and digits

Preoperative daytime splinting of the wrist in neutral for a period of 6 months is invaluable in improving the range of passive movement at the wrist, limiting the degree of lengthening or release required and correcting finger postures that are secondary to the flexed posture of the wrist.

In young children I prefer to use the FCR for wrist extension. In older children and adults preoperative assessment identifies those digits in which the FDP and FDS are contracting well, under moderate to good voluntary control. In these digits the FDS tendons can be used (Figs. 5 and 6). Both are frequently under good voluntary control and the muscle bellies are typically unscarred and healthy, even in adults. The FDS to the middle and ring are preferred for independence, strength and an excursion which is in excess of that required for wrist movements and digital movements. The ring FDS is transferred to the ECRB and the middle finger FDS to the extensor digitorum communis (EDC). The FDS transfer reduces the tightness of finger and wrist flexion with the result that lengthening procedures are not invariably required and ulnar deviation may be reduced. Mechanically, the individual FDS muscles are a little underpowered for wrist and finger extension. In practice the increased excursion available in the transfers appears to compensate for this.

When there is poor control of the FDP in a digit the FDS should not be used. Use of the index should be avoided if there is any prospect for pulp-to-pulp pinch grip. The superficialis minus finger is prone to develop a swan-neck deformity and any tendency for this prior to transfer warrants a FDS tenodesis at the time of harvest. Following transfer, it is important to appreciate that independent flexion will be reduced in the fingers. The index profundus is usually separate but the middle, ring and little finger FDP tendons have little independence.

Surgery commences with musculoaponeurotic lengthening of the flexor mass avoiding the FDS. Persisting tightness of the wrist and fingers is treated by musculotendinous lengthening of the FDP, FCU, palmaris longus (PL), flexor pollicis longus (FPL) and FCR. This usually allows passive composite digital and wrist extension. The FDS tendons are harvested at the AI pulley which has not been complicated with swan-neck deformities, but if these pre-exist, the finger is opened and a radial slip FDS tenodesis is made. The transfer is through the interosseous membrane. In my experience taking these transfers around the forearm does not sufficiently abolish their flexor action and can lead to persistent flexion contractures and failure. The FDS tendons are woven to the ECRB and EDC and tensioned to a maximum of 20° of wrist extension and at least 30° of passive wrist flexion. The fingers are tensioned to full MCP joint extension with the wrist extended.

Postoperative care. The goals of protection during the healing phase and early restoration of mobility and function without adhesion formation may seem at first glance mutually exclusive. Early mobilisation of flexor tendon repairs and more recently extensor tendon repairs has successfully challenged this view. The tendon weave used in transfers is stronger than most methods for the repair of tendon injuries and the forces in the postoperative extremity in cerebral palsy are reduced by the procedures carried out, pain and swelling. It seems inexplicable therefore that almost all published postoperative regimens commence with a period of 4 weeks of immobilisation. This can only lead to adhesions, with reduced excursion and power of the transfers. I have little doubt that many a good transfer has ended up as an immobile checkrein that has ultimately stretched and failed. My own preference is early mobilisation starting on the first postoperative day with the exception of biceps lengthening where a weave is not used. In 3 years of this regimen, I have not seen a single failure due to disruption of the transfer, but have frequently encountered previous transfers that were initially immobilised, fixed in scar tissue with no pull-through.

At surgery the patient is splinted in a maximum of 20° of wrist extension and extension of the MCP joints. Over-extension can precipitate flexor spasm that severely exacerbates postoperative pain. The following morning a new, removable splint is applied. Active and passive exercises are carried out every 2 h. Effective analgesia with or without antispasmodics is essential.

Joint procedures at the wrist

Wrist fusion is a means to stability in patients with severe spasticity associated with significant problems of hygiene, cosmesis and pain[15] or for stability in athetosis.[16] Splinting can be used to demonstrate the effect preoperatively. In the spastic group proximal row carpectomy with adjunctive soft tissue releases (e.g. wrist flexor tenotomy) may be necessary to achieve a neutral position.

Forearm rotation

The classic posture is one of forearm pronation with the deforming forces being the PT and pronator quadratus (PQ) muscles.

Wrist and digital procedures may improve rotation and should be carried out first if planned. The resting position of the limb following surgery for forearm rotation needs careful consideration of the individual patient's needs. If the volitional control of pronation is poor and active or stable pronation is lost after surgery, a patient could be severely compromised, for example if wheelchair bound.

Treatment is based on the Gschwind and Tonkin classification of the deformity into four groups.[16]

In group I, active supination is present beyond the neutral position and surgery is rarely required, although flexor aponeurotic lengthening used for the wrist and fingers may give rise to an improvement of 20° to 30° of supination if the PT is included.

Group 2 patients, in whom active supination is present to the neutral position or less, the authors found that release of PQ could be expected to improve the arc of supination by 40°. My own preference in this group is to reduce the strength of pronation by PT transfer, thus preserving active pronation by the PQ. The PT is rerouted converting its pull to supination. This transfer has been found to be more effective than a simple tenotomy.[16]

Group 3 patients, in whom no active supination is present but some passive range is maintained, PT transfer achieved an average arc of rotation of 97°.

Group 4 patients, in whom no active supination is present and passive range is restricted, some restoration of range can be carried out by flexor aponeurotic lengthening, but will often require release of the PQ. Combined PQ and PT release and transfer should be avoided, as this will abolish active pronation risking an incapacitating supination contracture.

Postoperative care

The forearm is classically immobilised for 4 weeks in 45° of supination with the elbow flexed to 90°. My practice is to splint the arm in supination and the elbow in as much extension as was obtainable at the time of release, immediately postoperatively. On the first postoperative day, the elbow component of the splint is kept in place, the remainder replaced with a removable splint and range of motion exercises commenced.

The thumb-in-palm deformity

The key elements are spasticity of the thumb ray flexors and adductors, weakness of the extensors and abductors, changes at the metacarpophalangeal joint, and secondary soft tissue tightness in the web space. The classification of House[4] forms a sound basis for treatment.

In Type I deformities there is a simple metacarpal adduction contracture causing a tight first web in the absence of tightness of the FPL. Correction is by release of the first dorsal interosseous and the adductor pollicis (AP). The web space skin is satisfactorily lengthened with a four-flap Z-plasty.

In type 2 deformity there is a simple metacarpal adduction contracture with a MCP joint flexion deformity due to additional spasticity of the flexor pollicis brevis (FPB). Correction is as above for type I, with additional release of the origin of the FPB.

In type 3 deformity there is a simple metacarpal adduction contracture combined with a MCP joint hyperextension deformity. Correction is as for type I with stabilisation of the MCP joint, however the MCP joint is often improved by correction of the wrist position.

In type 4 deformity there is a metacarpal adduction contracture combined with MCP and IP joint flexion and represents a type 2 deformity with additional FPL spasticity. Correction is as for type 2 with lengthening of the FPL. Tightness of the FPL may also be present in type 3 deformity.

Techniques

FPL lengthening. This is a combined musculotendinous and musculoaponeurotic lengthening (see above). In severe cases, step lengthening of the FPL tendon may be required.

First web and intrinsic release. In most cases, a 4-flap Z-plasty is used to gain access to the tissues and divide the fibrous bands that cross the tight web space. The first dorsal interosseous muscle is identified and the dense fascia on its dorsal surface is divided. The fascia overlying the adductor pollicis is similarly divided. In mild cases this may provide adequate release. In more severe cases the first dorsal interosseous and AP origins are released, with FPB and carpal tunnel release for resistant cases. In these circumstances where severe skin tightness is anticipated, the skin approach to the first web is longitudinal and a pedicled dorsoradial index finger flap used for resurfacing.

Rebalancing the thumb. Brand[9] has described the biomechanics of thumb movements in detail and the reader is referred to his text as an invaluable source of information for planning. Studies comparing the different approaches are lacking. The aim is to move the thumb ray out of the palm so as not to impede finger flexion and if possible, to allow grasp between the fingers and the thumb. Tonkin[16] uses proximal traction on the abductor pollicis longus (APL) and extensor pollicis brevis (EPB) to determine which tendon is most effective in achieving this aim. The chosen tendon is then augmented. As can be seen from Table 3, the FCR is a good transfer for the APL and the PL for the EPB. When these transfers are unsuitable or have been used for other purposes, alternatives are the FDS and brachioradialis.

House[4] reported good or excellent results in 36 of 56 patients, 42 of who underwent augmentations of the APL, EPB and EPL in various combinations using the PL, FCR and brachioradialis. Techniques to adjust the pull of the augmented APL to improve its vector for extension or abduction are described. The importance of MCP joint volar stability is emphasised with the need to fuse this joint to control hyperextension that cannot be balanced by tendon transfers. The authors reported a tendency of the brachioradialis transfer to form adhesions and used augmentation of the extensor pollicis longus (EPL) to correct persisting IP joint flexion deformity after appropriate lengthening of the FPL.

Matev[17] described successful correction of a thumb-in-palm deformity in 82% of 56 patients using an alternative approach of shortening of the APL and EPB. This study raised the important observation that the thumb ray extensors were stretched rather than truly weakened. This lent itself to the simpler approach of tightening these muscles rather than augmenting them, with the implications that this has on retraining after surgery. Finally, the results were achieved without the need for joint stabilising procedures or reinforcement of the EPL.

Joint stabilisation in the thumb. The commonest problem is hyperextension at the MCP joint. Failure to correct this by transfers can be treated by a MCP sesamoid capsulodesis. The radial sesamoid, which lies within the volar plate of the joint, is attached proximally to the metacarpal head–neck junction.

MCP joint arthrodesis may be necessary if capsulodesis fails to control hyperextension or there is a persistent flexion contracture resistant to releases and transfers.

This can be achieved by careful, sharp excision of the articular cartilage and Kirschner wire fixation of the joint. The epiphyseal plate is preserved using this technique and normal growth can be expected.[18]

CMC joint fusion is a last resort option for a metacarpal adduction contracture that cannot be controlled by other means. Interphalangeal joint fusion similarly is a rarely used option for instability at that joint when soft-tissue options have failed.

The fingers

Swan neck deformity

Swan neck deformity may develop from spasticity of the long extensor or from hyperextension of the digits in an attempt to overcome a flexed posture at the wrist.

Many cases of can be corrected by correcting the flexion deformity at the wrist and this should therefore precede surgery on the finger deformity. The surgical options are extensor tendon lengthening, volar translocation of the radial lateral band, volar plate capsulodesis and FDS tenodesis.[16]

Authors preferred technique of FDS tenodesis. The FDS radial slip is sutured through bone holes to the proximal phalanx tensioning the tenodesis at 5° of PIP joint flexion. (Fig. 7).

Postoperatively, most authors immobilise the proximal interphalangeal joint for a period of 2 weeks. In my own practice, patients are usually undergoing early active motion following wrist and hand tendon transfers. In these patients, gentle passive range of motion is commenced on the first postoperative day.

Intrinsic muscle spasticity

The fingers rest in a posture of flexion at the MCP joints and extension at the IP joints which may progress to swan neck deformity. Although considered a problem of the intrinsic muscles, some extrinsic (extensor) pull is often present. As a result correction of the flexed wrist position may be helpful. If the deformity persists and is preventing PIP joint flexion or opening of the hand, surgical lengthening can be achieved by the interosseous muscle slide. Through two dorsal longitudinal incisions over the index-middle and ring-little intermetacarpal spaces, a subperiosteal release of the dorsal and palmar muscles is carried out maintaining an extensor pull on the MCP joints.

The MCP joints are immobilised in extension for 4 weeks, before range of motion exercises are carried out. My own preference is for early active mobilisation with a resting splint that maintains MCP joint extension, in between exercises.

THE FUTURE

CP is the commonest cause of motor disability affecting the upper extremity in children and yet in our region in the United Kingdom only a fraction of patients are referred for surgery, often as adolescents or teenagers when deformity and habits are firmly established and difficult to treat successfully. Education of carers, charities, primary and secondary care physicians and their support workers is an essential step to increase general awareness of the potential benefits of treatment and to

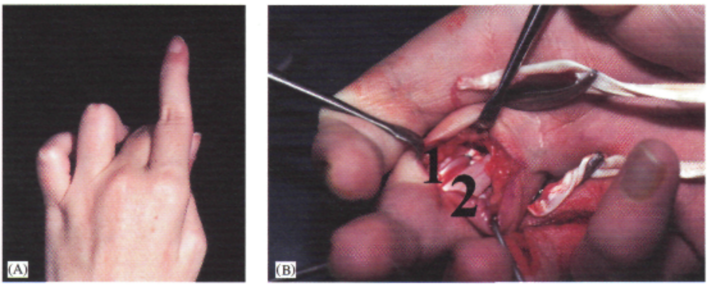


Figure 7 (A, B) Swan neck deformity and correction by FDS tenodesis (1 = radial slip of FDS used for tenodesis, 2=FDP).

encourage early referral. In parallel with this, multidisciplinary teams in specialist hand surgery units need to prospectively gather information and carry out research to fill the large gaps that exist in our knowledge of this condition. The views that the deformities seen in CP are inevitable and cannot be prevented or modified by timely intervention and the surgical techniques based on these views are being challenged. The goal should be a multidisciplinary team approach with early referral, function-preserving surgery and early mobilisation to maximise the benefit of that surgery.

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