Progression and Correction of Deformities in Adults with Cerebral Palsy

Mark Paterson is a paediatric orthopaedic consultant in London with a particular interest in cerebral palsy and neurodisability. He is Secretary to the British Society for Surgery in Cerebral Palsy and is the Orthopaedic Representative for Scope.

Introduction

Cerebral palsy (CP) is a lifelong condition, and although the neurological lesion may be static, the musculoskeletal sequelae change significantly throughout life.

The effect of spasticity upon the musculoskeletal system is summarised in Figure 1.

The childhood years are spent in a constant attempt to maintain functional muscle length and joint ranges as the skeleton grows. Management - stretches, serial casting, orthotics, myoneural blockade and muscle-tendon surgery - reflects this ongoing battle against threatened muscle contracture and consequent joint deformity.

In adult life, growth is clearly no longer a deforming force. However, it is replaced by a number of other potential general problems, such as stiffness, weakness and fatigue. These will affect individuals to a varying extent depending on their level of disability. Furthermore, there are specific problems related to individual joint segments that are again associated with particular levels of disability. These general and specific features will be described below, together with strategies for coping with them.

With the exception of hip reconstruction and spinal surgery, there is nothing technically difficult about the actual operative procedures described. The skill, such as it is, lies in the assessment of these patients and their problems and in determining an appropriate plan of management. It follows that one should refer to an orthopaedic surgeon with an interest in cerebral palsy who will be sensitive to the needs and priorities of their patients, who will be able to analyse movement, and who will have close links with physiotherapists, occupational therapists and seating specialists.

Spectrum of disability

The spectrum of disability following skeletal maturity is very wide. Any consideration of orthopaedic problems in the adult with CP has to take into account the following groups:

1. walking adults leading independent lives
2. obligatory chair users
3. severely involved adults requiring constant care
4. a small number of adults with isolated upper limb problems.

Some adults with mild spastic diplegia will have few or no problems with daily life as an adult. More severely affected adults will experience increasing problems as the abnormal muscle forces take their toll on the joints of the limbs and on the spine.

1. Spasticity – the sequence of events.

The walking adult with CP

Weakness and fatigue

The importance of muscle weakness is often not recognised. In cerebral palsy, the fundamental problem is one of muscle imbalance, but although the deforming muscle group may be relatively powerful compared with its antagonists, its absolute power is often significantly lower than normal[1,2]. In the presence of spasticity or fixed contracture it may be difficult to demonstrate weakness. Indeed, there are some CP patients who depend on their spasticity to maintain an upright position. The fact that their muscles may be weak often comes as a surprise to the young adult. There is an understandable tendency for the owners of these troublesome tight muscles to assume that because they are stiff and resisting stretch, they must be strong muscles, but in many cases, nothing could be further from the truth. In practice, this situation tends to be well tolerated by the teenager and young adult, but often by the late twenties and early thirties, there is a definite reduction in walking distance and complaints of fatigue on carrying out tasks previously performed with ease[3]. In many ways this is similar to the experience of those with lower motor neurone disease such as polio or spina bifida. There is little the orthopaedic surgeon can do here in terms of intervention, but a full explanation of the phenomenon is invaluable, even if the message is hard to take.

Stiffness

Loss of movement range is a common problem once CP patients move away from the care of paediatric physiotherapists and enter a busy independent life on their own. The reduction in availability of physiotherapy in adult life is dramatic and appears to be a universal problem[4]. Much can be done by the patients themselves, but some joint

2. A. Marked valgus deformity causing pain on medial border of foot
B. Lateral radiograph showing subtalar fusion.

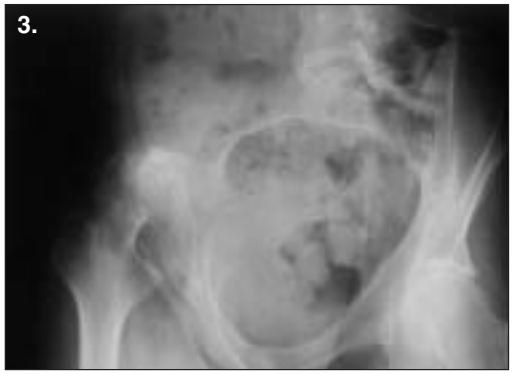
ranging exercises do require help from another individual, and there is undoubtedly the "personal trainer" factor of encouragement and motivation. Although the literature is split on the value of stretching and strengthening exercises in adult CP[5,6] my experience is that patients do benefit from short intensive "top-up" blocks of physiotherapy once or possibly twice a year. The best advice that can be given to the diplegic patient leading an independent life is to sign on with a gym (preferably one with a pool). It is often possible to find someone in these places who is willing to help out with a few hamstring stretches once a week.

There is probably little role for muscle-tendon lengthening surgery in the adult diplegic patient because of the risk of creating unacceptable weakness. Use of tone-reduction techniques such as myoneural blockade with botulinum A or ethanol also have limited effect because the contractures in adults are largely structural in nature rather than dynamic. One exception to this is the patient (usually hemiplegic) with a dystonic component to their motor disorder, as these often respond well to botulinum.

Specific problems

Persistent flexed knee gait is the commonest pattern in the adult walking patient and represents a continuing imbalance between a weak quadriceps mechanism and more powerful and contracted hamstrings. This crouch position may have been exacerbated by inappropriate heel cord lengthening in childhood. It will also be worsened by increased body weight. Patellofemoral compression forces in the weight-bearing flexed knee are high, and can give rise to painful degenerative arthritis in relatively early adult life. Contracture of the quadriceps can give rise to an additional complication at the knee in the form of a high-riding patella (patella alta). Loss of the normal articular relationship with the femur may further exacerbate discomfort. This is a very difficult problem to manage and there is no easy surgical solution. Lengthening of the hamstrings will serve no useful purpose if the quadriceps are too weak to maintain knee extension. Repositioning of the high patella in its normal groove involves lengthening the quadriceps mechanism and reefing the stretched patellar tendon, and runs the risk of further weakening an already compromised quadriceps mechanism. One alternative may be to avoid excessive soft tissue surgery and instead perform a shortening extension femoral osteotomy with reefing of the patellar tendon. The problem is far from solved.

Low back pain is very common in the walking adult with CP. This is purely mechanical in nature and occurs as a result of the excessive pelvic movements which are characteristic in this condition. Treatment is symptomatic.



3. Neglected but painless dislocation of the right hip in a 51 year old man.

Foot deformities established in childhood tend to become more troublesome in adult life. These may be simple hallux valgus and claw toe deformities or they may represent more fundamental hindfoot valgus or varus malalignments. Many minor problems can be dealt with adequately by appropriate in-shoe orthotics, but surgical correction should be seriously considered to re-establish a comfortable and stable base for walking (figure 2).

In late adult life, the problems of degenerative arthritic change become dominant. There is no reason why someone with spastic diplegia or hemiplegia should not be offered a total hip replacement. It used to be said that there was an unacceptably high risk of implant dislocation due to the spasticity, but more recent studies have shown that this is not the case[7].

The obligatory chair-user with CP

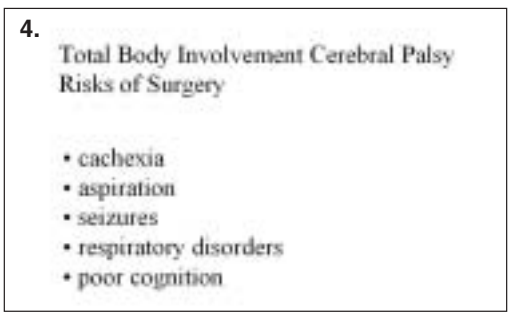
These patients will either be more severely affected diplegic patients who have lost useful walking function or, more commonly, patients with total body involvement CP.

The orthopaedic priorities in the non-walking patient with total body involvement centre around the spine and pelvis. The aim is to achieve and maintain symmetry as far as possible. Loss of symmetry resulting from, say, unilateral hip dislocation and the associated pelvic obliquity, leads inexorably to seating problems and will exacerbate any pre-existing scoliosis.

The management of the subluxing and dislocating hip will be dealt with in the next section on the severely involved adult requiring constant care.

Scoliosis may continue to deteriorate well into adult life[8]. The best opportunity to correct and stabilise this surgically has been missed by the time patients reach skeletal maturity. Bracing with a spinal orthosis is neither practical nor effective in the majority of adults with spinal deformity. Thus, the importance of good specialist seating cannot be overstated. Without such seating, a patient with an unbalanced spinal curve will not be able to use his or her upper limbs to their optimum effect. They will either be slumped to one side or will be using one hand constantly as a support to prevent such a posture. It is unrealistic to expect such seating to achieve any corrective function; this is accommodative seating designed to support in a functional position.

Occasionally the scoliosis is so severe that there is intractable pain from impingement of the lower ribs on the iliac crest. There is also a risk of decubitus ulceration. By this stage surgical correction may then be necessary, but the risks of surgery are high and the degree of correction achieved is likely to be disappointing.



4. Total body involvement cerebral palsy – risks of surgery.

There is much discussion between orthopaedic and spinal surgeons over the respective roles of hip subluxation and scoliosis in the genesis of the ultimate deformity, and which to correct first. Some maintain that the scoliosis is the primary deforming factor, causing pelvic obliquity and encouraging the uncovering of the femoral head, whereas others take the opposite view, stating that the dislocation and pelvic obliquity cause the spinal curve to develop. It is a somewhat sterile and unhelpful argument, and it is probably more useful simply to regard the spinal and hip deformities as interactive, and to take steps to prevent these deformities developing in the first place.

There is a tendency to ignore hamstring contractures in the obligatory chair-user as there is no walking ability that requires knee extension. This is a mistake. The hamstrings not only flex the knees but due to their origin from the pelvis also pull on the pelvis at the back. Hamstring contractures therefore result in a posterior tilt of the pelvis so that the patient is sitting on their sacrum rather than their ischial tuberosities. Not only does this create problems with pressure areas, but it gives rise to a compensatory thoracic kyphosis which is frequently marked and very disabling. This is one situation where soft tissue surgery in adults with CP can be extremely beneficial. Hamstring lengthening will result in a more upright spinal posture and easier and more comfortable sitting.

The severely affected adult with total body involvement CP

The major orthopaedic problem in this group is that of hip dislocation (figure 3). Subluxation of one or both hips may continue to progress through growth into adult life. This may be a silent event, but it is likely that we underestimate the level of discomfort and disability associated with dislocated hips[9]. There may be problems with cleaning and dressing because of the associated adduction and flexion contracture. As discussed above, a unilateral dislocation will result in asymmetric sitting and lying posture, and exacerbation of any pre-existing scoliosis. Occasionally, in an apparently painless bilateral dislocation symmetry may be maintained and there may be no sequelae in terms of seating, lying or care problems.

However, surgery in this group of patients is a major undertaking as can be seen from figure 4. Perioperative mortality rates for hip reconstruction surgery may be as high as 4%[10]. If the dislocation is apparently asymptomatic, it is tempting to leave well alone.

If pain, posture or caring difficulties conspire to make hip surgery necessary, a decision has to be made with respect to the condition of the femoral head. If the progression of subluxation has been brisk, the femoral head may still be in good condition and it is reasonable to plan a full reconstruction of the hip joint. This will involve soft tissue releases of the adductors and hip flexor muscles, osteotomy of the proximal femur to correct rotational and valgus deformity, and possibly also a pelvic osteotomy to realign or reshape the acetabular roof (figure 5). If however the head has been out of the acetabulum for a long time, the articular surface will have become worn and ulcerated from pressure against the capsule of the joint. If such a hip is reconstructed to produce a reduced femoral head, it may continue to be painful due to this articular damage. In the adult patient this is likely to be the case, and under these circumstances, an alternative to reducing the dislocated head is required.

Longstanding painful hip dislocations in the adult may

5. A. Preoperative radiograph showing dislocated left hip
B. Postoperative radiograph after soft tissue releases and both femoral and pelvic osteotomies

6. A. Failed surgery for recurrent spastic dislocation of left hip
B. Radiograph showing good abduction and level pelvis following proximal femoral resection.

7. 18 year old lad who was turned down at countless job interviews because of the appearance of his upper limb. He had little useful function in this limb and simply wanted it out of the way. Tendon lengthening at the elbow and wrist enabled him to keep his hand in a pocket and prevent excessive posturing. He got a job six weeks after surgery.

be treated by proximal femoral excision. In this procedure, the upper third of the femur is removed and the muscles around the hip fashioned into a cushion between the femoral bone end and the empty acetabulum (figure 6). This is a salvage procedure that, if performed correctly, gives a good chance of providing a comfortable and mobile "hip"[11]. It is not indicated for anyone who relies on bearing weight to effect transfers, as no guarantee can be given that this would be possible after such surgery. The procedure has suffered from a bad press in the past, largely because it has not been executed properly. It is sometimes confused with the Girdlestone procedure in which only the femoral head and neck are removed. In CP this is an inadequate resection and will lead to poor results. The formation of heterotopic bone may also impair the outcome, but this can be minimised by the administration of either preoperative radiation or postoperative indomethacin. One should expect 70-80% success rate from this procedure in terms of pain relief and improvement in ease of care.

Upper limb problems in the adult with CP

In a younger age group, carefully selected patients will achieve improved function from tendon transfer and tendon lengthening surgery. In the adult patient it is unlikely that surgical intervention in the upper limb will improve function, with the single exception of wrist fusion for the athetoid or dystonic extremity[12]. In this situation painful posturing can prevent otherwise reasonably useful function, for example, switch control or keyboard use.

Surgery is more likely to be indicated for cosmetic reasons or in extreme deformity to improve access and hygiene (figure 7). The severely involved patient may have severe thumb-in-palm or clenched-fist deformities that lead to skin maceration and infection. Simple wrist and finger flexor tenotomies can make a huge difference to the patient's and carer's life.

Summary

It will be seen that there is relatively limited scope for the orthopaedic surgeon to address established deformities in adult life, and nothing that can be done to prevent the natural history of weakness and fatigue. It is therefore essential that every opportunity is taken during the growing years to ensure that everything has been done to prevent deformity so that the patient with cerebral palsy, regardless of level of disability, faces the onset of adult life with optimised function.

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Correspondence to:
J Mark H Paterson FRCS
consultant orthopaedic surgeon
Barts and the London NHS Trust
E-Mail. jmhp@blueyonder.co.uk