INTRODUCTION

Total body involvement cerebral palsy (TBICP), also known as spastic quadriplegia, is the most severe end of the spectrum of disease that is cerebral palsy (CP). Children with this condition have a multitude of problems, and their orthopaedic problems, though important, are only one component of their condition.

The patient with CP has functional priorities that may be at odds with those perceived by the treating orthopaedic surgeon. The children and their parents are most concerned with the following, in this order:

1. Communication
2. Activities of daily living
3. Mobility in the environment
4. Walking

Whereas children with hemiplegia and diplegia will usually walk, are often of normal intelligence and may function relatively well in the community, patients with TBICP are likely to be completely dependent on their care providers. They are less likely to be of normal intelligence, and communication with them can be difficult. They often have difficulty feeding themselves, and may require a feeding gastrostomy to allow adequate nutrition. Their swallowing difficulties may lead to aspiration and subsequent recurrent chest infection, producing still more debilitation and malnutrition, and this can affect their ability to withstand, and to recover from, surgery.

It can be extremely difficult to communicate with children with TBICP making diagnosis very exacting. With the most important indication for hip surgery in this group being pain or its prevention, it is important that the patients can connect with their environment, as otherwise extensive and potentially life-threatening hip or spine surgery may be of little value.

In treating the child with TBICP, we need to address ourselves to treating the facets of the condition that impact on the child most, and maximally improve quality of life.

A child with TBICP is likely to communicate best with its carers in the upright position, i.e. in an appropriate wheelchair (Fig. 1). To this end, surgery should be directed at the reduction of pelvic obliquity and spinal deformity, by the correction of severe scoliosis and hip dislocation. Communication will also be improved if the child is comfortable, so a painful, dislocated hip merits treatment. Activities of daily living are easier if the child may be seated rather than lying, and perineal hygiene will be facilitated by improving abduction of the hips.

Mobility is likely to involve the use of the wheelchair (Fig. 2) and comfortable seating may require surgical correction of equinus deformities of the ankle. This may also facilitate transfer from bed to chair, allowing the parents to manage transfers in a home setting rather than an institutional one.

All of the above are important parts of a much bigger picture — the management of a child with TBICP requires the input of a wide range of professionals preferably as a multidisciplinary team working together with the family. These include the general practitioner, the paediatrician,
physiotherapist, occupational therapist, orthotist, nurse specialist, social worker, speech therapist and dietician.

**NATURAL HISTORY OF TBICP**

CP is defined as a non-progressive disorder of movement caused by malfunctioning of, or damage to, areas of the brain which control motor function. Although the brain injury or abnormality which causes it is non-progressive, the effects of it on the child are not. As the child grows, the asymmetry of muscle pull produces deformities of the joints both in the axial and the non-axial skeleton. The deformities will tend to leave the child more difficult to seat and to care for and should therefore, if possible, be prevented or minimised and once present need attending to so as to facilitate the best possible quality of life for both the patient and their carers.

As the majority of deformity develops during growth, once the child reaches adulthood, the requirement for orthopaedic intervention decreases, allowing significantly decreased surveillance after skeletal maturity.

Deformity in CP passes through three phases and the management varies depending on which phase the patient’s disease is in:

1. Correctable, dynamic deformity (treatable with physiotherapy and orthotic management).
2. Fixed deformity (soft-tissue surgery ± bony surgery).
3. Fixed deformity with bony involvement (bony and soft-tissue surgery required).

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**Figure 1** Child with spastic quadriplegia in custom-built wheelchair. Despite a significant scoliosis, she is comfortable sitting almost upright in this chair.

**Figure 2** Custom-made wheelchair for a child with TBICP. Note the moulding of the seat and the thoracic pads to support the child despite her scoliosis.

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82 CURRENT ORTHOPAEDICS
SPECIFIC JOINTS

Hip

The hip is the joint in which most orthopaedic management of the totally involved child takes place. There is asymmetric muscle tone with most specifically the abductors, flexors and internal rotators being involved. This tends to place the hip in the most unstable position (think of the position in which the majority of dislocated arthroplasties occur) and in conjunction with increased femoral anteversion and valgus of the neck leads to subluxation and eventually dislocation of the hip. The time of maximum risk is between 4 and 12 and it is at this time that surveillance needs to be greatest. Early management may delay or abort dislocation, whereas treatment of an established dislocation is more complex and the results less predictable (Fig. 3). The prevalence of established hip dislocation by adolescence is in the range of 25–40%.

There is not only asymmetry of the muscles, but also asymmetry of the two sides of the body. This, with the asymmetric muscle pull of the spinal muscles results in scoliosis and pelvic obliquity. The final result of these two can be the windswept deformity, where one hip is abducted and the other adducted, and treatment needs to be tailored as appropriate (Fig. 4).

Knee

Flexion contractures of the knee develop due to spasticity of the hamstring muscles, particularly the medial hamstrings. Severe knee flexion contractures may lead to the child sitting on the sacrum rather than the buttocks and cause pressure sores and significant lumbar kyphosis, making percutaneous hamstring lengthening occasionally necessary.

Ankle/foot

The gastrosoleus muscle complex is the dominant influence around the foot, along with the tibialis posterior. This will result in an equinus deformity at the ankle, which is initially dynamic and eventually becomes fixed. The tibialis posterior produces inversion, so that an equino-cavo-varus deformity of the foot is also a common pattern of deformity.

Spine

Scoliosis is a frequent problem with the incidence increasing with the severity of the condition. Between 60 and 74% of individuals with spastic quadriplegia will have scoliosis. Neuromuscular scoliosis is resistant to brace treatment and, particularly in TBI, is more likely to progress after maturity with a relatively smaller curve than in patients with idiopathic scoliosis. A curve of 50° or more is likely to progress especially if the patient is bedridden and has a thoracolumbar or lumbar curve (Fig. 5). Treatment for the more severely affected patient who retains some connection with their environment, apart from expert fitting and manufacture of a custom-built wheelchair, may well therefore involve spinal surgery.

Upper extremity

In the upper extremity, there is flexion and pronation at the elbow and flexion at the wrist and fingers. This is initially dynamic, but may become fixed as the child grows. Indications for upper limb surgery in the child with CP are limited, even more so in the totally involved child, and realistically the only indications in the child with TBI are to improve use of a computer keyboard or communication board or enhance hygiene in a severely contracted hand.
Ashipsubluxationandscoliosisaresocommoninthe
totally involved patient, careful surveillance is important
during growth. Observation is both clinically to observe
range of motion of the hips and curvature of the spine,
and radiologically after the age of 4 to monitor the posi-
tioning of the femoral head and the degree of dysplasia of
the acetabulum. Radiographs of the spine are
indicated when clinical evidence of scoliosis becomes
apparent.

Assessment of the hip joint is by the use of the
Reimer’s index (a percentage assessment of the amount
of the femoral head lying lateral to the rim of the
acetabulum—an increasing Reimer’s index over time
suggests subluxation); the acetabular index (the angle
made by the roof of the acetabulum and Hilgenreiner’s
line, which transects the two triradiate cartilages); and
an assessment of whether Shenton’s line is broken
(Fig. 6).

Figure 4  (A and B) Clinical photograph and radiograph show-
ing classical windblown position. Note abducted hip on the right
and adducted, dislocated hip on the left.

Figure 5  (A and B) Radiographs showing TBICP scoliosis.
These are typically either a long thoracolumbar curve or a long
C-shaped curve.

MANAGEMENT OF THE CHILD WITH
TOTALLY INVOLVED CEREBRAL
PALSY

As hip subluxation and scoliosis are so common in the to-
tally involved patient, careful surveillance is important
Reimers index should be less than 25% at age 4. Greater than this is abnormal and an index of > 50% will not resolve spontaneously, with approximately one-third going on to dislocation. An acetabular index of > 30° is also suggestive of a hip with a significant risk of dislocation (Fig. 6).

**Early management**

Early management is through stretching exercises, physiotherapy and bracing. The purpose is to maintain range of motion, and to prevent contractures.

At this stage the use of botulinum toxin (Botox) temporarily defunctions the spastic musculature and may allow range of motion to be regained or improved. This is particularly useful for the spastic hamstrings, adductors and gastrocnemius, and to a lesser extent in the upper extremity. Recent literature has reported its use in the iliopsoas.

Baclofen may also be used to decrease spasticity in TBICP. Its use orally is associated with side-effects, particularly drowsiness, and it is more effective given through an intrathecal pump. This however can only be used in larger children with the body mass to accommodate the pump and is associated with other complications such as infection and migration of the catheter.

As the child becomes older, the dynamic deformities will become fixed. It is preferable to delay this for as long as possible as simple stretching and bracing will no longer provide adequate range of motion. Once the contractures become established, soft-tissue surgery may be used to allow movement to be regained before joint dislocation and bony deformity occur. This is particularly useful in the treatment of equinus deformity with gastrocnemius recession (transverse division of the tendon in the muscle belly, leaving the muscle intact) or, in the more severely affected child, with simple tenotomy of the Achilles tendon. Hamstring tightness may also be treated with tenotomy of the medial (and variably the lateral) hamstrings to allow knee extension to be regained.

Opinions vary as to the management of early subluxation or the hip at risk of subluxation. The use of soft-tissue releases delays the onset of hip subluxation and results up to 6 years are satisfactory but it does not correct the underlying bony abnormality and the soft-tissue contractures tend to recur with time. For these reasons, the senior author is of the opinion that soft-tissue surgery alone only ‘delays the inevitable’ and that soft-tissue releases such as iliopsoas recession and adductor release (tenotomy of the adductor longus and brevis and gracilis at the origin from the pelvis) are only effective in the long term when combined with bony surgery.

**Bony surgery**

The aim of surgery around the hip is to prevent subluxation, or to reduce a dislocated femoral head in later cases.

The indications for surgery are twofold:

1. to alleviate pain;
2. to allow comfortable sitting.

Pain is a common complaint in TBICP, with an estimated prevalence of 50% in patients with a dislocation. A severely subluxed femoral head will develop an indentation on the dorsal aspect due to pressure from the overlying capsule and the spastic abductors and medial notching from the acetabular rim and the ligamentum teres. These combine to produce a triangular shape to the dorsal aspect of the femoral head. This gives rise to a stiff painful articulation that will not permit comfortable sitting or perineal hygiene and this is the reason to intervene surgically, in order to either prevent dislocation or to treat the established condition.

Surgical treatment of an early or moderate subluxation involves soft-tissue release to allow the femoral head to be placed in the acetabulum, combined with a varus derotation osteotomy of the femur to produce the stability required to maintain that reduction. This procedure externally rotates the distal fragment to correct the excessive femoral anteverision and places the neck in varus to correct the valgus deformity.

More severe degrees of dislocation require shortening of the femur in combination with the varus and derotation, and may also require a pelvic osteotomy to provide adequate coverage of the femoral head. As the acetabular deficiency tends to be posterior (as opposed to anterior in DDH) the pelvic osteotomy used is an acetabuloplasty leaving the anteromedial cortex of the ilium.
intact. This allows lateral and posterior cover, rather than the anterior cover provided by a Salter innominate osteotomy (Fig. 7).

**Windswept hips**

Treatment of the patient with windswept deformity of the hips requires careful assessment and release of the muscles involved in creating the deformity (adductors, flexors and medial hamstrings on the internally rotated side, and abductors, iliobial band and lateral hamstrings on the externally rotated side), along with varus derotation shortening osteotomies of the femur to produce more stable hip anatomy. The deformity is prone to recurrence following surgery.

**Salvage surgery**

In the older patients with a painful deformed femoral head, operative reduction may not be an option. Relief of pain in these patients is likely to require a salvage procedure. Various procedures have been described, which include:

1. valgus osteotomy (to increase abduction and relieve pain — results inconsistent);
2. resection of the proximal femur (results poor);
3. interposition arthroplasty in combination with resection of the proximal femur (results better but risk of heterotopic ossification);
4. arthrodesis (less useful in the more severely involved patients);
5. total joint arthroplasty (useful in the less severely involved quadruplegics, providing good pain relief but necessitating a period of spica treatment post-operatively to prevent dislocation).

If possible, dislocation should be prevented, as the results of early treatment are generally better than those of salvage surgery. This having been said, recent literature suggests that the outcome can be satisfactory in more severely affected adults with quadriplegic CP, with only 30% of adults with dislocations being in pain and the results of resection/interposition arthroplasty being satisfactory in this group.

**Spinal surgery**

Spinal surgery is indicated for patients with curves at risk of progression either before or after skeletal maturity. The goals of surgery are to prevent progression, improve truncal balance and sitting posture and prevent the pulmonary and skin complications of a large curve. Surgery is likely to consist of posterior instrumentation and fusion, from upper thoracic to lower lumbar (Fig. 8), and will include the pelvis if there is evidence of significant pelvic obliquity (which is generally the case). If the curve is very large and stiff, anterior release may also be required, preferably at the same surgical episode or 1–2 weeks earlier with interval halo-femoral traction.

**CONCLUSIONS**

Treatment of the patient with TBICP is best undertaken with a multidisciplinary team involved in managing all aspects of their care. From an orthopaedic point of view, prevention is better than cure and depends on the stage the patient is at in terms of the correctability of the deformity.
The majority of orthopaedic treatment is likely to be concerned with the hip and with the spine, and to a lesser degree with the ankle and foot. The decision to operate must be taken with all care providers, with the degree of pain and the patient’s ability to connect with their environment being the factors of greatest importance.

**FURTHER READING**

**NATURAL HISTORY**


**RESULTS OF SOFT-TISSUE RELEASES**


**RESULTS OF VARUS OSTEOTOMY**


**RESULTS OF COMBINED FEMORAL AND PELVIC OSTEOTOMY**


**RESULTS OF SCOLIOSIS SURGERY**


Figure 8  (A) Thoracolumbar curve, preop. Note that the left bending film shows the ribs impinging on the pelvis, and also demonstrates the pelvic obliquity. This was treated with a ‘unitrod’ fixation (B), using Luque sublaminar wires and Galveston fixation into the pelvis.