

CURRENT TOPIC

Therapeutic choices in the locomotor management of the child with cerebral palsy—more luck than judgement?

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In the past 15 years or so our understanding of many aspects of cerebral palsy has significantly broadened. Traditional concepts regarding aetiology have been radically challenged, and we now acknowledge that “birth asphyxia”, once felt to be implicated in the majority of cases, has a causative role in approximately 10–15% of cases.¹ Antenatal factors are recognised as having a predominant aetiological role; newer techniques in neuroimaging, progress in exploring homeobox genes, and other developments, have supported this view.^{2,3}

Therapeutic nihilism, once prevalent in this field, has fortunately receded with the development of an array of new antiepileptic drugs for children with refractory seizures (seen so often in the child with cerebral palsy), and the enthusiastic application of new feeding techniques has done much to improve the nutritional status and well being of these children.⁴

Newer techniques have also been employed in the management of locomotor impairment and although the benefits conferred by certain treatments are clear, other management options are more controversial. A variety of treatments have been developed in recent years; in addition to conventional orthopaedic surgery and physiotherapy (usually based on Bobath techniques in this country), multilevel surgery,^{5,6} intramuscular botulinum injections,⁷ selective dorsal rhizotomy (SDR),⁸ intrathecal baclofen (ITB),⁹ targeted training,¹⁰ and sophisticated orthoses¹¹ all have their advocates. Less orthodox strategies too, such as hyperbaric oxygen, cranial osteopathy, and lycra suits,¹² have also been in vogue. In short, confusion abounds.

Cerebral palsy is a heterogeneous condition; one may, therefore, legitimately argue that its management should be highly individualised. For professionals in the field, however, there is a plethora of choice, but a paucity of guidelines. The familiar leitmotif here is: Which child? Which treatment? Although there may be some debate concerning optimum treatment in the non-ambulatory child, the greatest pitfalls are created by the lack of established protocols for the walking child with spastic cerebral palsy.

In some parts of the country, an orthopaedic surgeon may have a particular interest in a certain procedure and apply this widely, whereas

in another area a different management protocol may be in favour (perhaps influenced by resource, or indeed lack of it). Which treatment the individual child eventually receives is thus often influenced more by factors such as post-code rather than what is clinically optimal. Furthermore, many children are allocated a treatment regimen without having a full gait analysis—yet such analyses often influence or change clinical perceptions.¹³ Even if resource and professional skills were evenly distributed however, treatment allocation would still be something of a lottery, as indications for different treatment modes, and criteria to be employed in their choice, are not well established.

Gait analysis for all children with cerebral palsy?

The physician needs objective criteria for analysing problems of locomotion in order to make rational decisions about surgery, orthotics, physiotherapy, and drug treatment. Three dimensional instrumented gait analysis improves clinical assessment of walking and often clarifies the reason for deformities in children with spastic cerebral palsy, especially in the rotational (transverse) plane.¹⁴ Simultaneous biomechanical analysis can be shown graphically of the coronal, sagittal, and transverse kinematics (joint ranges) in the gait cycle.

When combined with video film, coincidental dynamic electromyography, and assessment of the energy cost of walking, analysis is even more useful. Kinetics (resulting from combining movements and forces) provides knowledge of moments acting at joints in the lower limbs but can only be obtained if the child can make individual foot contact with a force platform embedded in the floor. Thus it is independent walkers who most benefit from gait analysis. Children with hemiplegia, diplegia, and mild quadriplegia can all be routinely assessed, but less able diplegics who require ambulatory aids and walk with high energy costs may benefit from individually tailored gait studies. Children with ataxic or dyskinetic cerebral palsy present particular challenges in gait analysis because of the risk of marker displacement and erratic gait pattern which overshoots the measuring space; these problems are not, however, insurmountable.

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To ensure success the child must be able to cooperate in simple barefoot walking tasks in the laboratory, wearing shorts and stick on markers. Size rather than age (because of marker size) may be a barrier to data collection, but with technology continuously improving, ever smaller children are being assessed; obesity may also produce problems with marker placement. We have carried out gait analysis on willing 3 year olds. In very young or cognitively impaired children we sometimes have to resort to video filming only. This compromise can be useful as shown in a recent study.¹⁵

Gait analysis has been perceived by many as a “research tool”,¹⁶ but as it is now apparent that gait analysis alters therapeutic decision making when compared with observational analysis, its place in routine practice is becoming accepted.^{13 17} Not only is it valuable in helping to determine intervention in many clinical situations, it is also a useful audit tool for recording outcome for a range of therapeutic options.¹⁸

Outcome measures remain a vexed question and there are few validated measures in this field. The continuing maturation of the CNS and growth of the child undoubtedly need to be taken into account in any outcome measure, and pre and post interventional gait analyses provide a valuable though expensive academic outcome tool.

Other less technical procedures have been developed in recent years as outcome measures, eg. the Gross Motor Function Measure (GMFM), Paediatric Evaluation of Disability Inventory (PEDI), the Functional Independent Measure for Children (WeeFIM), Gross Motor Performance Measure (GMPM), Activities Scale for Kids (ASK).^{19,20} These scales are used to measure changes in functional independence or impact on life style. Their main disadvantage is their lack of specificity as compensatory mechanisms may be involved in achieving better function, rather than the intervention under scrutiny. Their major advantage of course is that they do not require a gait laboratory and can be performed in the home or school by a trained therapist.

Conventional surgery

For many years the mainstay of surgical treatment for children with cerebral palsy was tendon lengthening, bony fusions and derotation osteotomy. These procedures have traditionally been carried out one at a time and have led to the recognition of a “birthday syndrome” (orthopaedic surgery each year followed by physiotherapy to relearn a walking technique, and a further operation the next year to release the consequent contracted muscles).²¹ Such programmes have done little for the child’s self-esteem, his socialisation or his education and are gradually being replaced by a multi-level surgical approach.

Multilevel surgery

The results of orthopaedic surgery in cerebral palsy have previously been unpredictable, some children responding poorly to an operation which has benefited others with a similar gait.

Surgeons now acknowledge the illogicality of addressing single components of what is an extensive and complex disorder, and with the help of gait studies can differentiate between primary and compensatory changes in patients. A comprehensive surgical prescription should be based on the best evidence available from both clinical examination and gait analysis, to avoid irrevocable surgical treatment of a “compensation”. We have shown in our own unit that multilevel surgery is a good option in the hemiplegic or diplegic child where cognition and emotional maturation are adequate to comply with postoperative rehabilitation.⁶ This surgery should be undertaken at the start of the adolescent growth spurt to allow “fine tuning” by nature. Until then, the child should be treated with physiotherapy, orthotics, and possibly temporary muscle paralysis. Although this approach is well accepted in orthopaedic circles, there is nevertheless a need for its evaluation by means of a randomised controlled trial.¹⁷

Botulinum toxin

The use of intramuscular botulinum toxin to block neuromuscular transmission in spastic muscles has become commonplace in recent years.^{7 22} Its prime use has been in calf muscles to overcome dynamic equinus of the ankle in children with hemiplegia or diplegia. The effects of botulinum toxin are transitory and injections must be repeated every three to four months. The technique is simple, appears to be safe, and successfully paralyses muscle, but whether it benefits gait in the long term is unproven. Evidence of a beneficial effect has not consistently been found in clinical trials.²³

The indications for single level injections require careful scrutiny in a condition where the whole limb is affected; a more holistic approach is, however, emerging in the use of botulinum toxin.²⁴ Multilevel treatment is effective in the younger child where there are no fixed contractures, where gait analysis is available to help with precise identification of target muscle, and where high quality orthotic provision is available during rehabilitation.

Botulinum toxin has many niche applications. It has an expanding role in planning surgical intervention in both upper and lower limbs.²⁵ It protects after tendon transfer, especially during healing of muscle working over joints. It is also important in facilitating orthotic use in the pursuit of an overall postural goal.

Selective dorsal rhizotomy

Disinhibition of the spinal reflex arc resulting from an upper motor neurone lesion is thought to be the basis of spasticity in the child with cerebral palsy. Selectively dividing portions of the dorsal lumbosacral roots of the spinal cord, and thus interrupting the spinal reflex arc on the sensory side leads to reduction in spasticity without causing paralysis.

Selective dorsal rhizotomy (SDR) has been explored and greatly refined in the past 20 years; it is regarded as an important treatment

option for children with cerebral palsy in Canada, the USA, and Australia.²⁶ It has, however, been almost universally shunned in the UK.²⁷

All agree that the procedure reduces spasticity, but whether it confers functional benefit is controversial.²⁸ Two recent randomised controlled trials showed that SDR was beneficial,^{29,30} although a third study showed that intensive physiotherapy was as effective if assessment was carried out one year postoperatively.³¹ The debate has been fuelled by lack of standardised selection criteria, lack of agreement on intraoperative techniques, and no uniform outcome measures. Despite its popularity in the New World, there have been few long term studies on the outcome of SDR. Those which have been published (although not controlled studies) suggest that children who have had SDR early, have a favourable long term outcome with respect to laboratory and functional assessment.^{8,32} The review of our first 15 carefully selected diplegics and quadriplegics who have had this procedure is encouraging (Cole, Patrick, and Roberts; manuscript in preparation).

Intrathecal baclofen

Unlike the oral preparation, baclofen delivered intrathecally (ITB) sidesteps the blood-brain barrier; this accounts for its greater efficacy. It migrates into superficial layers of the spinal cord where essentially it substitutes for insufficient levels of GABA, and so directly reduces spasticity. It has been used in the USA for children with cerebral palsy for at least 10 years, but to date it has found little favour in this country and our own experience with it has been limited. Only 70–80% of children with cerebral spasticity respond to ITB, thus trial doses of bolus intrathecal injections are a necessary screening procedure.³³ Children whose spasticity is diminished by such means may be suitable candidates for long term continuous infusion of the drug by a programmable subcutaneous pump. The major advantages of such a treatment are that (a) it is reversible, and (b) the therapeutic response can be titrated; however, infection and catheter related problems requiring surgical correction (migration, kinking) are problematic and the sheer size of the subcutaneous pump may preclude its use in smaller children.

Intrathecal baclofen has been shown to be beneficial in children with a notable dystonic element (although not in other dyskinetic forms of cerebral palsy).⁹

Some American workers feel that ITB is useful in walking diplegics, but the 10% risk of serious complications makes us less enthusiastic.³³ Furthermore, apart from its reversibility we believe ITB has no advantage over conventional multilevel surgery. In our view the proper use of ITB is in the non-mobile child with severe spasticity where, apart from moulded seats and oral muscle relaxants, previously there has been little therapeutic solace available. The judicious use of ITB now offers greater comfort by reducing spasticity and ultimately bony deformity. It also facilitates easier

nursing, thus improving the quality of life of these unfortunate children and their often careworn parents.

Sophisticated orthoses

Abnormalities of body posture both between body segments and with respect to gravity are common in cerebral palsy. The use of orthoses broadly addresses these aspects by the application of external force to correct the relation between segments, between the body and gravity, or both.

A common use of orthoses in cerebral palsy is for controlling ankle position. However, in the walking child, the dynamic situation needs to be considered if the orthotic prescription is to be optimised. Where the ankle is in eversion during stance, the relation between the shank and the ground reaction force may lead to a flexing moment about the knee and often the hip. The simple application of a rigid orthosis does not necessarily address the postural abnormality, and it is the application of gait laboratory tuning which produces sophistication in the orthosis rather than its material or design features.¹¹ Tone reducing orthoses have been proposed; however, the evidence that they work by the purported mechanism (protuberances on the sole) is tenuous.³⁴

Contracture correction devices (CCDs) use orthotic principles to apply a stretching force across a joint that is controllable and consistent across a range of positions. Used as an adjunct to botulinum toxin or soft tissue surgery, CCDs allow safer and more effective treatments for flexion contractures than aggressive surgical release that is often accompanied by unwelcome neuropraxia of unpredictable duration.³⁵ After surgery, CCDs may prevent recurrence of contracture as a result of continuing skeletal growth.

Targeted training

This promising treatment technique is based on the principle that the normal child achieves motor control in a cephalad to caudad direction. It uses specially designed equipment to provide the correct level of support so that the child can learn to control one or two joints at a time, rather than being overwhelmed by too many control demands. After careful assessment, the highest body segment lacking in control is targeted (often the head in children with cerebral palsy); by using the programme for some period every day, it is possible to progress motor control learning in a downwards direction. Encouraging results are being achieved with the technique.¹⁰

Less orthodox treatments

There are always new treatments appearing for the child with cerebral palsy, for example, hyperbaric oxygen, cranial osteopathy, reflexology, etc. Many of these fail to stand both the test of time and scientific scrutiny. While such “trends” are at their height, however, we need to be well informed about them so that we can properly advise vulnerable parents. It is important to keep an open mind on these issues as

some of them (for example, lycra suits) do appear to provide some help, although the scientific premises for their use are not always clear.¹²

Conclusions

- Therapeutic dilemma is inevitable in this area of increasing treatment options but in which there is a dearth of guidelines. Evidence of the value of some techniques is flimsy, the ways in which we compare results in many cases have not been validated, there are no acceptable outcome measures in general use, and the whole area is bedevilled by more than a healthy splash of professional prejudice and scepticism.
- There is still much to be discovered in the basic science of neuromuscular function. The phenomena of tone, spasticity, etc. are still poorly understood and it is perhaps only when we have a clear idea of these neurophysiological concepts that questions such as “which child?”, “which treatment?” will be answered with confidence.
- A basic “rule of thumb” has emerged in the therapeutic approach to the child with cerebral palsy and there is now a quickening tempo and enthusiasm to rationalise management. A severely impaired child needs effective orthopaedic supervision to prevent spinal curvature and hip dislocation. Such inter-related deformities are eminently preventable, either by ITB or surgical means, yet sadly many children are denied this chance.

For the walking child with spastic cerebral palsy the objective is to counter or minimise the negative long term effect of spasticity and weakness during the period of growth. Simple measures of stretching, walking, and orthotic use can be supplemented by oral muscle relaxants and particularly botulinum toxin. These “bridging” measures augment function, keep the child’s joints straight, and preserve walking at a reasonable energy cost. Some children will go on to require more major intervention and SDR needs to be considered where spasticity is gross but where power and control are relatively intact. Such cases are not common and the “best buy” treatment for children requiring major intervention is likely to remain multi-level orthopaedic surgery.
- Professional objectivity in this field can nevertheless be improved and there appear to be two major stepping stones to achieving this. Firstly, strengthening the multidisciplinary approach to assessment of the child with a complex gait disorder and acknowledging the role of the bioengineer, neurophysiologist, and mechanical engineer as well as the paediatrician, orthopaedic surgeon, paediatric neurologist, and physiotherapist. Secondly, the many phenotypes of cerebral palsy make collaboration between different centres essential if meaningful numbers in different study groups are to be achieved. The recent consensus between UK and European centres to establish a

network of registers and agree on the classification of cerebral palsy is undoubtedly a major contribution to establishing such multicentre studies.³⁶

- The careful selection of children, the choice of the most effective, safe treatment, and an objective evaluation of functional outcome, must be our goal. Defining and refining selection criteria in respect of the multiple treatment options available for the child with cerebral palsy would seem no small step in the opening decade of this new millennium.

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