

**Dystonia Europe 20<sup>th</sup> Anniversary Conference**  
**“Achievement and Hope”**  
**Edinburgh 2013**

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Over the last 20 years there has been a growing perception that dystonia is under-recognised in children and that dystonia has strong developmental origins involving basal ganglia, cerebral cortex and cerebellum.

Edinburgh is one of the paediatric centres with a long tradition of attempting to characterise movement disorders in children, recognising existing limitations of our understanding of the mechanisms of hypertonus and measure the contributions of dystonia and spasticity to movement patterns in children. It is therefore fitting that the Dystonia Europe 20 year celebrations, reflecting on what has changed in our treatment of dystonia in children should take place in Edinburgh.

TTS Ingram was a pre-eminent Edinburgh clinician who made a major contribution to our understanding of cerebral palsy and like his successor, J Keith Brown was deeply influenced by the work of Denny Brown and the growing interest in the functions of the basal ganglia.

The first half of the 20th century saw the rise of orthotics and braces to attempt control of body alignment and function in children with movement disorders. Yet none of these methods have been shown to prevent fixed deformity.

Consequently, it has been claimed that the second half of the 20th century was the ‘orthopaedic era’ of management of cerebral palsy, culminating in the development of gait analysis laboratories to help in ‘planning’ for multi-level orthopaedic procedure, often involving bilateral femoral and tibial osteotomies and muscle lengthening procedures, requiring simultaneous adjustment of 8-12 muscles chiefly for the ‘spastic phenotype’ due to the recognition that orthopaedic procedures are less predictable and thus less successful in children with dystonia or dystonic chorea.

Selective dorsal rhizotomy has been hailed as very successful for children with spastic diplegia, but not dystonia. However many centres are quietly abandoning the procedure because of the profound permanent weakness which the procedure uncovers.

These orthopaedic and neuro-ablative surgical procedures have, at the very least, required increasingly careful evaluation of the nature of the underlying movement disorder, measurements of function and quality of life.

Botulinum toxin became a very important tool for relieving muscle spasms in the early 1990s but although heralded originally as a means of preventing contracture in 'spastic' (probably hyperextensible) mice, randomized controlled trials of botulinum toxin injections with and without splints or orthoses have not demonstrated prevention of contracture in children with cerebral palsy.

Deep brain stimulation (DBS) has come at an interesting time in this field when all these 'older methods' of management have been found wanting.

The singular success of DBS for genetic dystonias has led to a renewed interest in the motor neurophysiology of the brain and concepts of refining motor focus with high frequency DBS. Defining new targets for DBS may help a larger number of children affected by cerebral palsy. Thus a convergence of strategies designed for very selected motor disorders may be relevant for a wider group of disorders.

This should be interpreted in the context that recovering lost motor skills is quite a different process from developing previously non-existent motor function.

Other promising avenues are the effective management of pain in dystonic disorders and managing the neuroinflammatory component of encephalitis which produces receptor antibodies often associated with severe movement disorders.

The real challenge to these earlier palliative strategies is to harness advances in neuroimaging, clinical genetics, neuropharmacology and neurosurgery to improve our early therapeutic strategies for children with movement disorders dominated by dystonia with the aim of altering an otherwise grim and unremitting natural history.

Perhaps we can look forward to the 21st century as the neurophysiological and neurosurgical era of management of dystonia in children.