

Management of Spastic Cerebral Palsy in the UAE: An Overview



Khalid A Mohamed
FRCPCH,

is a Consultant Paediatric Neurologist at Sheikh Khalifa Medical City, Abu Dhabi. He graduated in Khartoum and trained in the UK, with training in General Paediatrics. He trained in paediatric neurology in Liverpool and Oxford. Dr Mohamed was also formerly an honorary clinical lecturer at the University of Liverpool and the school of Tropical Medicine, and a consultant Paediatric Neurologist in Liverpool for 5 years. Dr Mohamed's main areas of interest include epilepsy and neuron-metabolic disorders.



Sabahat A Wasti,

is a Consultant in Physical Medicine and Rehabilitation. Having completed his Specialist Training in Rehabilitation Medicine in Leeds, he became Consultant in The Sheffield Teaching Hospitals Trust in 1998. He moved to Sheikh Khalifa Medical City Abu Dhabi in July 2007. He has first hand experience of Rehabilitation Services Development both in the UK and the UAE.

Correspondence to:

Khalid A Mohamed, FRCPCH,
Consultant Paediatric Neurologist,
Sheikh Khalifa Medical City,
Abu Dhabi UAE.
Email: kmohmed@skmc.gov.ae

Cerebral palsy (CP) is a diverse group of disorders caused by a static injury to the developing brain between conception and 3 years of postnatal life. CP is common, affecting roughly around 3 in a 1000, and its prevalence and aetiology is different in different parts of the world. Most children with CP will have spasticity as the main motor disorder and it can be classified either according to which body areas are affected: hemiplegia, diplegia, tetraplegia, or the movement disorder type: spastic, athetoid, ataxic and hypotonic cerebral palsy.

Most children with cerebral palsy are initially hypotonic. With time, tone changes and spasticity patterns begin to emerge and become manifest. In many children, spasticity interferes with their functional abilities and, when severe, soft tissue contractures and skeletal deformities may develop. In some extreme cases joint disarticulation and spinal malalignment results in subluxation or total dislocation of joints such as hips and / or scoliosis and kyphosis. In a minority, spasticity is useful and contributes to patients functional capabilities providing much needed strength. This is particularly true for stability provided through spasticity around the knees that in turn assists mobility.

The natural history and impact of spasticity is variable, ranging from mild functional limitation to severe disability. In many children CP can be progressive with symptom exacerbations corresponding to the periods of linear growth.

The prevalence of cerebral palsy in the United Arab Emirates

The population of the UAE is heterogeneous, comprising of nationals and expatriates. Children make up 50% of the local population. There are no available studies to indicate the prevalence of CP in the UAE. Our practice experience indicates that it is probably similar to the western populations, with some exceptions; for instance there is likely to be a higher incidence of inherited causes such as genetic microcephaly and other syndromes associated with CP, perhaps due to consanguinity and close relation marriages.¹ There is also a notable increase in multiple births and prematurity related to fertility therapy. Obstetric and neonatal care is generally well delivered and, without firm figures, perinatally acquired CP is probably comparable to

the other established healthcare systems. Post natal causes for CP are relatively uncommon and include central nervous system infections in infancy and traumatic and non traumatic brain injury.

Management of spasticity:

Spasticity management requires an individualised and coordinated team effort to deliver good and efficient care; if provided well, spasticity care can improve function and reduce complications such as deformities and contractures. It may prove cost effective because of the reduction in repeated hospital admissions and reduced number of orthopaedic procedures and other interventions required. There is also the benefit of the functional gain in the affected patients. International groups have analysed the cost effectiveness of therapies used to treat spastic CP and concluded that management of spasticity is at least cost neutral if not cost effective.² In the UAE the provision of such a service is still in a state of evolution.

Standards of care for spasticity management:

It is internationally agreed that children with cerebral palsy require regular ongoing physical therapy;³ this should ideally be delivered by well trained qualified paediatric neuro-physiotherapists. All CP patients require a regular follow-up by a developmental paediatrician and regular review by a paediatric neurologist, to evaluate the changing needs related to growth and the need to make necessary adjustments to the management plan in conjunction with the physical therapist.

Some patients will require more specialised interventions, such as posture management through specialised seating systems, botulinum toxin therapy or intrathecal baclofen pump. Alongside developmental paediatricians and physiotherapists, such patients must be reviewed by a whole team including specialised paediatric neurologists, occupational therapists and rehabilitation engineers. When necessary, paediatric orthopedic surgeons and neurosurgeons should be consulted.

The role of physiotherapy has been formally evaluated in CP while newer therapies such as botulinum toxin and intrathecal baclofen have

been extensively studied. Randomised trials and systematic reviews have confirmed the short term benefit from the use of botulinum toxin.⁴ Studies have, however, failed to demonstrate a persisting long term benefit. There are some long term studies that support the role of botulinum therapy in conjunction with other techniques such as abduction devices and serial casts. Botulinum toxin has also been demonstrated to reduce the risk of hip dislocation in children with CP.⁵ Intrathecal baclofen delivered via a pump is gaining popularity as an effective and longer term therapy for severe lower limb spasticity.⁶ These therapies require specialised expertise and should only be delivered by a trained team. They are costly and require thoughtful resource allocation and should ideally be delivered in tertiary centres.

Resource allocation and health insurance:

The UAE has recently adopted an insurance based health care system. This has introduced a western style, market place health-care model. The insurance companies are charged by the health care providers. Patients or the employing organisation are directly charged if the insurance cover is lacking. Although private medical care has always thrived in the UAE, the new system has replaced a largely state funded system. However, healthcare provision for children with disability is still mostly state funded even for the ex-patriate population; the UK model of school health and provision of specialised care in special schools is only available in some of the state funded humanitarian projects that provide schooling and essential therapies for children with disabilities. Several non-profit organisations and private institutes also exist throughout the country and provide physical therapy, occupational therapy and speech therapy. Across the Emirates, however, service provision for children with cerebral palsy is inconsistent with some areas being better resourced than others.

The provision for spasticity management for children with CP is mainly in the large tertiary centres, such as Sheikh Khalifa Medical City in Abu Dhabi. There are four paediatric neurologists in the UAE, who provide consultations and medical treatment for spasticity. Two are trained to give botulinum toxin therapy. Whilst there are very few specialised paediatric neurophysiotherapists and occupational therapists in Abu Dhabi, children who receive the botulinum toxin therapy have access to this expertise. One hospital has a visiting developmental paediatrician who provides occasional botulinum toxin treatments.

Botulinum toxin therapy:

Botulinum toxin is well known to improve spasticity in the short term. It is now widely used for spasticity and offers best results in children with focal spasticity that is severe enough to affect function or produce symptoms or complications; an excellent review by

Boyd et al¹ concluded unequivocal short term benefit from this therapy. It is now agreed that short term goals can be clearly identified and influenced by botulinum toxin. Botulinum therapy is safe and produces few and tolerable side effects. Recent studies questioned the role of this therapy in the long-term outcome of CP and spasticity. The author has researched and co-authored work⁷ studying the long term effects of botulinum toxin type A. Using the Gross Motor Function Measure (GMFM) and Paediatric Disability Index (Pedi) we concluded that outcome measures may not be sensitive enough to detect the functional and symptomatic impact of botulinum therapy and a Cochrane review by our group has further confirmed the lack of evidence for long-term benefit.⁸

There is a growing body of evidence that, apart from a minority of children receiving botulinum toxin for symptomatic relief, this therapy is not effective if not accompanied by an integrated therapy and orthotic programme.

In the UAE we have been providing regular botulinum toxin therapy mainly for three groups of patients:

1. Patients with severe adductor muscle spasticity considered at high risk of hip dislocation. In this group we currently have 5 patients. The hip alignment has been maintained and the children remain free from pain and discomfort due to either continued spasticity or partial or complete dislocation. We have been successful in preventing orthopaedic intervention over the treatment period of around 3 years.
2. A second group of patients with lower limb spasticity who are ambulant. Currently we have 15 patients under treatment. They demonstrate functional improvements with botulinum therapy followed by physiotherapy and orthotics, especially in those aged between 3-10 years old.
3. Children with troublesome spasticity that causes discomfort and hinders their daily care are being treated for symptomatic relief of pain and discomfort. Currently we have around 10 children who are receiving botulinum in this category. Our experience in these children is encouraging and the parental and carer satisfaction is reflected in the fact that a close link is maintained with our service for repeated injections and follow up.

Patients are referred usually from paediatricians and rehabilitation physicians as well as treating physiotherapists; patients are accepted from all areas of the UAE. Physiotherapy is often provided locally and the physiotherapist will facilitate communication with the orthotic and occupational therapy service. Close cooperation is more feasible with the therapists working in an institution than those working in the community. The service does not have access to motion analysis or formal gait evaluation, nor is there a specialised

upper limb service.

The service challenges within the UAE at the present time includes expansion and allocation of appropriate resources such as an integrated team of therapists and the availability of video equipment to undertake gait evaluations as well as access to formal motion analysis service.

Provision of a team approach to effectively manage children with upper limb spasticity requires the presence of a specialised upper limb physical and occupational therapist as well as the services of a hand surgeon; this model has been successfully developed previously by the author and colleagues in a large UK centre.

There is a large potential for further expansion and organisation of the service, especially given the current changes in the health service and the interest shown by various agencies in developing services for children with neurological disorders. Many new centres have been developed in the field of autism and learning disability, the field of physical disability is likely to follow suit and develop.

Intrathecal baclofen (ITB):

Intrathecal baclofen has been gaining popularity over the last 2 decades and the use of baclofen pumps is part of the standard care offered in many tertiary centres for children with severe spasticity. Delivery of baclofen to the intrathecal space produces dramatic improvement in the spasticity and reduces the risk of undesired central adverse effects that are known to limit the use of oral baclofen in the treatment of generalised spasticity.

The convenience of having an adjustable delivery system offers the potential for adjusting the dose depending on the patient's symptoms. Although many studies confirmed a benefit in walking patients with severe spasticity, non walkers with severe spasticity are the usual target group for ITB therapy. There is no service, to the author's knowledge, that offers ITB for children in the UAE although a small number of patients did receive this therapy abroad; none of those attends our tertiary spasticity service. Presently we are working with the neurosurgeons to set up this service.

Selective dorsal rhizotomy:

This therapy is particularly useful for children with severe diplegia where spasticity is the main barrier to walking. Dissection of carefully selected afferent nerve fibres at the spinal level breaks the reflex arc and reduces spasticity. Studies have demonstrated much improved spasticity and functional gains, however the procedure is very specialised and irreversible. The author is not aware of rhizotomy being undertaken anywhere in UAE.

Specialised seating service:

Historically the specialised seating service had been provided in the UAE via therapy

services and assessment conducted in conjunction with manufacturers. The author² is in the process of developing a multi-disciplinary seating assessment service. Around 10 patients have already been assessed in this relatively new service and parents and carers are being instructed in correct use of the equipment. The notion that systems are mobility devices is being discouraged and usage to improve and maintain biomechanics of sitting posture is explained and encouraged. It is too early to assess the effectiveness of this service.

Future projects

Healthcare is evolving in the UAE, the provision for children with cerebral palsy and other neurological disabilities will evolve and future developments are essential in order to deliver quality service for children with disability in general and children with spastic CP in particular. An integrated team approach and the provision of all spasticity management in one centre is likely to be the way forward for the tertiary care institutes. Furthermore, better access and facilities for children with CP within the school system will be required to ensure better function and participation in the community for children with disability in line with the recently approved World Health Organisation guidelines on this. ♦

REFERENCES

1. Al-Gazali UJ, Sztrihla L, Dawodu A, Bakir M, Varghese M, Varady E, Scorer J, Abdulrazzaq YM, Bener A, Padmanabhan R. *Pattern of central nervous system anomalies in a population with a high rate of consanguineous marriages.* Clin Genet. 1999 Feb;55(2):95-102.
2. Balkrishnan R, Camacho FT, Smith BP, Shilt JS, Jacks LK, Koman LA, Rascati KL, Mooney JF 3rd. *Cost impact of botulinum toxin use in Medicaid enrolled children with cerebral palsy.* J. South Orthop. Assoc. 2002;11(2): 71.
3. O'Neil ME, Fragala-Pinkham MA, Westcott SL, Martin K, Chiarello LA, Valvano J, Rose RU. *Physical therapy clinical management recommendations for children with cerebral palsy - spastic diplegia: achieving functional mobility outcomes.* Dev Med Child Neurol. 2008 Jun;50(6):450-5.
4. Boyd RN, Hays RM. *Current evidence for the use of botulinum toxin type A in the management of children with cerebral palsy: a systematic review.* Eur J Neurol. 2001 Nov;8 Suppl 5:1-20.
5. Graham HK, Boyd R, Carlin JB, Dobson F, Lowe K, Natrass G, Thomason P, Wolfe R, Reddihough D. *Does botulinum toxin a combined with bracing prevent hip displacement in children with cerebral palsy and "hips at risk"? A randomized, controlled trial.* J Bone Joint Surg Am. 2008 Jan;90(1):23-33.
6. Hoving MA, Evers SM, Ament AJ, van Raak EP, Vles JS; Dutch Child Spasticity. *Intrathecal baclofen therapy in children with intractable spastic cerebral palsy: a cost effective analysis.*
7. Mohamed KA, Moore AP, Rosenbloom L. *'Adverse events following repeated injections with botulinum toxin A in children with spasticity'.* Dev Med Child Neurol. 2001 Nov;43(11):791.
8. Moore AP, Ade-Hall RA, Smith CT, Rosenbloom L, Walsh HP, Mohamed K, Williamson PR. *Two-year placebo controlled trial of botulinum toxin A for leg spasticity in cerebral palsy.* Neurology. 2008 Jul 8;71(2):122-8.
9. Butler C, Campbell S. *Evidence of the effects of intrathecal baclofen for spastic and dystonic cerebral palsy. AACPDM Treatment Outcomes Committee Review Panel.* Dev Med Child Neurol. 2000 Sep;42(9):634.

The Association of British Neurologists Meeting

Liverpool: June 22 26th 2009

Message from Professor Alastair Compston, President of The Association of British Neurologists

The Association of British Neurologists is moving, at least for this year and next, to a single annual conference in place of the spring and autumn meetings that have been held for many years. However, we are also involved in one further meeting abroad in each of the next few years – The Netherlands (2010), Cuba (2011) and Boston (2012). The first annual UK-based conference will be in Liverpool from Monday 22 June to Friday 26 June. The venue is the spectacular new Arena and Convention Centre situated in the heart of Liverpool (European Capital of Culture in 2008) on the historic world heritage waterfront (www.acliverpool.com).



Alastair Compston, President.

High attendance is anticipated and the Association has set extremely competitive registration rates on the assumption that there will be a record number of delegates. For those who register early the cost is £350 for the whole week, including the reception and dinner, with 2 day and 1 day rates of \$275 and £150, respectively with half those costs for trainees and senior members. Registration will close on 10 June.

The programme is thematic in eight parts each having a scientific symposium and teaching course held in parallel with free communications and including one or more plenary lectures. In the session on stroke, Werner Hacke from Heidelberg will speak on Acute management. That is followed by an historical talk on Sir Charles Sherrington who spent many years in Liverpool given by Colin Blakemore – one of Sherrington's successors as Waynflete professor of physiology in Oxford. Richard Johnson, from John Hopkins University, will give the Gordon Holmes Lecture on Global hazards of infectious disease in the session on acute neurology, prion diseases and viral disorders. Angela Vincent from Oxford is the 2009 ABN Medallist and her lecture on Immunological disorders of ion channels is included in that part of the programme focusing on channelopathies, muscle

and mitochondrial disorders which is followed by a clinico-pathological conference presented by Andrew Lees from Queen Square. Eduardo Tolosa from Madrid will speak on Clinical and genetic diversity in Parkinson's disease in the session on neurodegeneration and movement disorders. The half-day devoted to dementia and behavioural neurology ends with a lecture by David Owen – In sickness and in power – in which he will argue that political decision-making in world

leaders may be contaminated by an acquired behavioural disorder manifesting as hubris. Finally, Jack Griffin, also from John Hopkins University, will give the Editors of Brain Lecture in memory of PK Thomas (on this occasion) on the Pathophysiology and mechanisms of peripheral neuropathy in the session on peripheral nerve disease. The meeting is interspersed with a symposium on academic neurology for trainees and a session highlighting free communications by younger members of the Association – Tomorrow's world - topped and tailed by an additional teaching course on neuro-ophthalmology and a practical session on how to survive as a neurologist.

The reception will be in the Merseyside Maritime Museum on the Albert Dock. The annual dinner is in the Liverpool Anglican Cathedral at which Baroness Ilora Finlay will be our guest and speaker. The evening will finish with a magnificent blast on the Cathedral Organ and a Son et lumiere performance.

All in all we hope that this much changed structure of the scientific meeting of the Association will appeal to all our members – young and old, NHS and academic appointees, and those wanting both continued professional development and glimpses into the future development of our rich and diverse discipline.

The organisers and Officers of the Association look forward to seeing you in Liverpool.



Werner Hacke



Colin Blakemore



Andrew Lees



Angela Vincent



John Griffin



Richard Johnson