SUMMARY OF NICE GUIDELINE 145 (JULY 2012) SPASTICITY IN CHILDREN AND YOUNG PEOPLE WITH NON-PROGRESSIVE BRAIN DISORDERS: MANAGEMENT OF SPASTICITY AND CO-EXISTING MOTOR DISORDERS AND THEIR EARLY MUSCULOSKELETAL COMPLICATIONS

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This guideline covers the management of spasticity and co-existing motor disorders and their early musculoskeletal complications for children and young people up to age 19 years with non-progressive brain disorders. The most common condition associated with this group is cerebral palsy. The guideline only covers the management of spasticity, and does not cover all aspects of the management of cerebral palsy.

Summary of recommendations
The guideline makes recommendations about the principles of care, relating to how care should be organised and delivered, and about interventions for management of the common problems associated with spasticity, its co-existing movement disorders, and its early musculoskeletal complications.

The guideline identifies key priorities for implementation, and these are *highlighted in this summary*.

1. Principles of care
Children and families should have access to a network of care, using agreed care pathways, supported by effective communication and integrated team working. A network team should comprise healthcare professionals who have expertise in paediatrics, nursing, physiotherapy, and occupational therapy, and who are experienced in the care of children and young people with spasticity. They should work in partnership with the child and family, and be unconstrained by professional and organisational boundaries.

Interventions from healthcare professionals outside the network team should be planned and discussed with the team, to ensure integrated care and effective subsequent management.

The network team should have a central role in the transition of care from children's to adult services.

A referral should be made without delay to appropriate members of the network team, who should *offer an individualised and goal-focused management programme that has been developed in partnership with the child, parents and carers.*

Age and developmentally appropriate goals should:
- focus on the body function and structure, activity, and participation domains of the World Health Organisation's International Classification of Functioning, Disability, and Health (children and youth version)
- be shared across the network team, and with other people involved in delivery of care

To help children and their parents to be partners in developing and implementing the management programme, offer appropriate and relevant information, with regular opportunities for discussion, about the potential for development, and the possible effect of the various treatment options.

There should be timely access to the equipment needed to implement management programmes such as sitting, sleeping and standing.

The child's condition should be monitored for response to treatment, worsening spasticity, and developing secondary consequences of spasticity, or to decide whether goals need to be changed.

There should be a pathway to monitor hips at risk of displacement, to include hip X-ray at age 24 months for all children with bilateral cerebral palsy; repeated annually for children at Gross Motor Function Classification System level III, IV or V, (Palisano et al, 2008), and 6-monthly for children with progressing hip migration greater than 30%.

2. Physical Therapy
There should be a *prompt assessment by a physiotherapist and, where necessary, an occupational therapist*, informing an individual tailored programme, with specific goals. It is important to take account of the views, needs and impact on the child and family when deciding on the content of the programme, and who should deliver it. The programme and equipment should be incorporated into daily activities, and should take into account any increased risks associated with secondary or associated health problems.

Specific strategies to consider include:
- 24-hour postural management, to prevent or

delay deformity, and enhance ability to participate in age appropriate activities. Appropriate strategies to consider include low load active and passive stretch activities, and training should be offered in how to deliver the programme.
- Interventions that involve task-focused active use, such as temporary restraint of affected arm to encourage use of other arm, or unrestrained use of both arms to enhance manual skills, delivered as a short (4-8 weeks) intensive programme.
- Muscle strengthening therapy for weakness, using progressive, repetitive exercises against resistance.

An adapted physical therapy programme is an essential component of management following botulinum toxin A injection, intrathecal baclofen pump insertion, orthopaedic surgery, or selective dorsal rhizotomy, and children and parents/carers should be made aware of this.

Undertake re-assessment at regular intervals to ensure achievement of goals and appropriateness of the programme.

3. Orthoses

Decisions about the use of orthoses should be based on the individual needs of the child, aimed at specific goals, and the balance of possible benefits and risks should be discussed with the child and their parents/carers. The orthosis should be correctly fitted, and information should be provided about how to apply it, and when/for how long it should be worn.

Specific recommendations are made about different types of orthoses for upper limbs, lower limbs and trunk, including elbow gaiters, rigid wrist orthoses, dynamic orthoses, ankle-foot orthoses, and body orthoses. The network team should review the orthosis at each contact with the child to ensure that it remains appropriate, is being used as recommended, and is not causing adverse effects.

4. Oral drugs

Oral diazepam should be used when rapid relief is required for pain, spasm or functional disability, starting with a bedtime dose, and increasing, or adding a daytime dose if the response is unsatisfactory. For sustained long-term effect, the use of oral baclofen should be considered. A low dose of oral baclofen should be commenced, increasing stepwise over around 4 weeks until optimum effect is achieved.

5. Botulinum Toxin type A

Consideration should be given to the use of Botulinum Toxin type A where focal spasticity or dystonia is causing problems with fine or gross motor function, care or hygiene, tolerance of other interventions such as orthoses or positioning equipment, or is causing pain or sleep disturbance.

Botulinum Toxin type A treatment is also suggested for difficulties resulting from rapid-onset spasticity following an acquired non-progressive brain injury.

Use caution where there is generalised spasticity, fixed contractures or bony deformity, a bleeding disorder, of where there may be problems with engaging with a post-injection therapy programme. It should not be used where there is severe muscle weakness, previous adverse reaction to Botulinum Toxin type A, or the child is receiving aminoglycoside treatment.

A baseline assessment of muscle tone, range of movement and motor function should be performed, involving a physiotherapist or occupational therapist, before deciding to treat with Botulinum Toxin type A. Follow-up assessments are suggested at 6-12 weeks, and again at 12-26 weeks after injection to inform the decision about further injections. Healthcare professionals from the network team who carry out the assessments and treatment should have expertise in child neurology and musculoskeletal anatomy.

The child and family should be given information about goals; what is involved in the treatment, including the assessment and injection process; any other interventions (e.g. orthoses, serial casting) that may be required; and the possibility of any adverse effects and how to recognise these.

The process of delivering the injections is considered, including the possibility of injecting more than one muscle, the use of topical anaesthetic and sedation, ultrasound guidance or electrical muscle stimulation, and when post-injection interventions such as serial casting and orthoses might be appropriate.

6. Intrathecal Baclofen

Continuous pump-administered intrathecal baclofen should be considered where, despite the use of non-invasive treatments, spasticity or dystonia is causing pain, muscle spasms, or postural, functional or care difficulties. Children who benefit are most likely to be bilaterally affected in upper and lower limbs, with motor function problems at GMFCS levels III IV or V. The guideline discusses contraindications, as well as the risks, benefits, and possible complications of the procedure.

Detailed recommendations are made regarding the testing procedure to assess the effects before pump insertion, including an appropriate pre- and post-test assessment covering those aspects relevant to the treatment goals, and their response to the test, including the views of the parents and child. The assessments and procedures should be carried out in a specialist neurosurgical centre.

The guideline also details what information and support should be provided to parents and children before and following pump insertion.

7. Orthopaedic Surgery

Orthopaedic surgery should be considered as part of the management programme for children with spasticity. *Assessment by an orthopaedic surgeon from the network team should be performed where clinical or radiological findings indicate that the hip may be displaced, or there may be spinal deformity*. Other reasons for an orthopaedic assessment include limited limb function, hygiene problems, or concerns about cosmetic appearance caused by contracture or bony deformity.

The network team should discuss the goals, risks, and post-surgery rehabilitation programme before undertaking orthopaedic surgery. The surgery should take place in a paediatric setting, carried out by surgeons from the network team who are expert in the concepts and techniques for this group of patients.

A thorough pre-operative assessment, including gait analysis where appropriate, should be carried out, and where several surgical procedures are involved to improve gait, these should be performed at the same time if possible. Outcomes of surgery to improve gait should be assessed 1-2 years later.

8. Selective Dorsal Rhizotomy

Selective dorsal rhizotomy (SDR) should be considered for children with spasticity at GMFCS level II or III. Teams offering SDR should participate in a national programme collecting specified outcomes on all patients assessed for SDR, whether or not SDR is performed.

Research recommendations

The guideline discusses the importance of some specific research questions:

1 What are the greatest inhibitors of functional ability in children and young people with upper motor neurone lesions?
2 What is the optimal postural management programme using a standing frame in children aged 1-3 years?
3 What is the clinical and cost effectiveness of Botulinum Toxin type A when used routinely or according to clinical need in children and young people at GMFCS levels I, II and III?
4 What is the clinical and cost effectiveness of intrathecal baclofen compared with usual care for children and young people at GMFCS levels IV and V?
5 Does SDR followed by intensive rehabilitation performed between ages 3 and 9 years for children at GMFCS levels II and III result in good community mobility as an adult?

This summary covers the main recommendations made in the guideline; for the full guidance document, and supporting information and implementation tools, the link given in the reference section below should be used.

Main Reference:

Spasticity in children and young people with non-progressive brain disorders: management of spasticity and co-existing motor disorders and their early musculoskeletal complications.
National Collaborating Centre for Women's and Children's Health. July 2012. http://guidance.nice.org.uk/CG145

Other references:

Palisano RJ, Rosenbaum PL, Bartlett D, Livingston MH (2008). Content validity of the expanded and revised gross motor function classification system. *Dev Med Child Neurol*, 50: 744-50