The Medical Management of Cerebral Palsy-Paediatricians’ Perspective

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What is Cerebral Palsy?
Cerebral palsy is a broad term describing a group of permanent disorders of the development of movement and posture causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbance of sensation, perception, cognition, communication, and/or behaviour, and/or seizure disorder.

Magnitude of the Problem in Hong Kong
According to a local cross-sectional survey of children aged 6 to 12 years performed between September 2003 and June 2004, among 435,572 mainstream primary schools and special needs school children, 578 were identified with cerebral palsy. The overall prevalence was 1.3 per 1000 children. The age-specific prevalence rate varies from 1.04 to 1.50 per 1000 children. The figures are compatible with other studies that range from 1.2 to 2.5 per 1000 live births.

Approximately 38% of children with cerebral palsy attended mainstream schools. For those studying in special schools, 96% attended institutions for the physically handicapped or the severely mentally handicapped. Among the 219 children with cerebral palsy studying in mainstream schools, 26% received educational support, and 61% received outpatient therapy. Only 12% received both supporting services. No educational or therapy support was received by 26% of children.

Risk Factors
There are prenatal, perinatal and post-natal risk factors for development of cerebral palsy. Certain risk factors could help us make an early diagnosis. The prenatal risk factors include intrauterine infections, malformations of cortical development, premature delivery. The earlier the maturity, the higher percentage of survivors would likely have cerebral palsy. The perinatal risk factors include: amnionitis, antepartum haemorrhage, neonatal meningitis, perinatal stroke and birth asphyxia. The post-natal risk factors include: CNS infection, ischaemic and haemorrhagic strokes, traumatic brain injury, hypoxic-ischaemic events etc. It is increasingly apparent that cerebral palsy can result from the interaction of multiple risk factors, and in many cases no identifiable cause can be found.

How to Make a Clinical Diagnosis?
Very often the diagnosis is delayed until the obvious clinical sign or deformity is detected. In order to make an early diagnosis, the recognition of early signs is important. Early signs include: developmental delay especially gross motor aspect (for example: poor head control, delayed sitting, standing and walking), toe walking, abnormal muscle tones either hypertonia or hypotonia, hyperreflexia, persistent primitive reflex, unusual posturing, early hand preference, spine deformity, failure to thrive, visual, hearing and speech problems etc.

What Other Types of Investigation Help to Make a Diagnosis?
Until recently, correlation between radiographic findings and clinical presentation in cerebral palsy was weak. Advances both in imaging technology and in quantitative motor assessments are now changing this.
picture. In the clinical context of cerebral palsy, MRI brain could reveal the following radiological features: 1) signal or cystic changes over periventricular white matter e.g. periventricular leukomalacia (fig.1&2) in ex-preemie diplegic patients; 2) signal changes over thalamus and basal ganglia in dystonic term babies with perinatal hypoxic-ischaemic injury; 3) features suggestive of cortical infarctions over watershed area, Porencephaly and Schizencephaly in hemiplegia; 4) involvement of the internal capsule and cerebral cortex in spastic tetraplegia; 5) features of malformation of cortical development.

When to Consider Other Differential Diagnoses Apart from Cerebral Palsy?

When a patient has spasticity or dystonia in the absence of any risk factors for cerebral palsy, one should consider other differentials. For example hereditary spastic paraplegia in a diplegic patient with family history of lower limb spasticity; adrenal leucodystrophies in a clumsy hemiparetic child with preceding normal motor function, cerebellar tumour or Pelizaeus-Mezbacher Syndrome in an ataxic child with nystagmus. Dopa responsive dystonia, tyrosine hydroxylase deficiency or other neuro transmitter deficiency should also be considered in a dystonic child without any perinatal risk factors. Many rare neuro-metabolic and neuro-genetic diseases could mimic cerebral palsy. These diagnoses require a high index of clinical suspicion, sometimes with the collaboration of chemical pathology and clinical genetics for laboratory confirmation.

Accompanying Impairments

Apart from motor impairment, there are many non-motor neurodevelopment or sensory problems associated with cerebral palsy that include: Intellectual Impairment, learning disabilities, epilepsy/abnormal EEG, speech and language disorders, psychological impairments, hearing impairments, sensory impairments, ophthalmologic defects, strabismus, dysfunctional voiding patterns, gastro-oesophageal reflux disorder, constipation, malnutrition, oral motor dysfunction, oral health problems, bone and mineral density disorders and sleep disturbances etc.

UMN Syndrome and Spasticity

Features of the UMN syndrome in cerebral palsy include both “positive signs” and “negative signs”. “Positive signs” include spasticity, spasms, clonus and hyperreflexia. “Negative signs” include weakness, loss of voluntary motor control, dexterity or fatigability. Botulinum toxin is useful in targeting the positive signs of UMN syndrome, while the negative signs are more difficult to manage. Spasticity has been most commonly defined as a velocity-dependent increase in passive muscle tone, associated with features of the UMN syndrome. It is seen in over 70% of those with cerebral palsy.

Why should We Make an Early Diagnosis?

The increase of the muscle tone will make muscle stiffer, causing muscle contracture. The latter will alter normal biomechanics and affect normal bony lever-arm system. Mal-alignment and skeletal deformity form a viscous cycle that can be broken with early effective intervention of the spasticity.

Classification of Cerebral Palsy

The definition of cerebral palsy covers a wide range of clinical presentations and variable activity limitations; it is therefore useful to further categorise individuals into classes or groups. The traditional classification schemes have been focusing primarily on the topography of the affected limbs, with an added modifier describing the predominant type of tone or movement abnormality. Based on dominant movement disorder and limb involvement, cerebral palsy can be classified into the following main groups: spastic hemiplegia, spastic diplegia, spastic tetraplegia, dyskinetic and ataxic cerebral palsy.

What is Dystonia?

Dystonia is a movement disorder in which involuntary muscle contractions (sustained or intermittent) result in twisting and repetitive movements. Examples include choreo-athetosis, tongue protrusion, facial grimacing and /or abnormal postures such as back arching. Dystonia often co-exists with spasticity and when present needs to be taken into consideration in planning spasticity treatments, as the outcome may be less predictable.

What is Botulinum Toxin-A?
Toxin-A
Possible Side-effects of Botulinum spasticity limiting arm abduction or causing pain. pronator spasticity limiting supination; shoulder opening or leading to excoriated skin flexor creases; grasping objects; fisting hand causing difficulty in hand Elbow flexor spasticity limiting arm extension, for functional use and ease of care in upper limb flexion contracture.

Foot deformity; causing crouching gait or difficulty with positioning or with perineal hygiene; Hamstring muscle spasticity or difficulty with splint use; hip adductor muscle Calf muscle spasticity causing equinus during walking or difficulty with perineal hygiene; Hamstring muscle spasticity causing scissoring during walking or difficulty with perineal hygiene; Hamstring muscle spasticity causing crouching gait or difficulty with positioning or sitting; tibialis posterior muscle causing equinovarus foot deformity; ilioptosas muscle spasticity causing hip flexion contracture.

For functional use and ease of care in lower limbs Elbow flexor spasticity limiting arm extension, for example for reaching, and affecting cosmetic appearance when walking; Thumb in palm causing difficulty in grasping objects; fist hand causing difficulty in hand opening or leading to excoriated skin flexor creases; pronator spasticity limiting supination; shoulder spasticity limiting arm abduction or causing pain.

How to Use Botulinum Toxin-A Injection?
The toxin is available in fixed dose ampoules and will normally require reconstitution in normal saline. The reconstituted solution is then injected directly into the spastic muscle. The toxin will spread within the muscle and cause local relaxation over two to three days. This relaxation effect usually lasts around 2-3 months.

Who may Benefit from Botulinum Toxin-A?
Botulinum toxin-A would benefit patients whose spasticity interferes with function or daily cares or causing pain, and in whom the muscles to be injected have some dynamic ranges as determined using the Tardieu Scale. The dynamic contracture is the difference between the R1 (first catch as the muscle is moved quickly through its range of motion) and the R2 (end of passive range of motion). The greater the dynamic range, the greater the potentials to have a favourable response to Botulinum toxin-A.

Clinical Indications of Botulinum Toxin-A Injection for Spasticity in Cerebral Palsy
For functional use and ease of care in lower limbs
Calf muscle spasticity causing equinus during walking or difficulty with splint use; hip adductor muscle spasticity causing scissoring during walking or difficulty with perineal hygiene; Hamstring muscle spasticity causing crouching gait or difficulty with positioning or sitting; tibialis posterior muscle causing equinovarus foot deformity; ilioptosas muscle spasticity causing hip flexion contracture.

For functional use and ease of care in upper limb
Elbow flexor spasticity limiting arm extension, for example for reaching, and affecting cosmetic appearance when walking; Thumb in palm causing difficulty in grasping objects; fist hand causing difficulty in hand opening or leading to excoriated skin flexor creases; pronator spasticity limiting supination; shoulder spasticity limiting arm abduction or causing pain.

Possible Side-effects of Botulinum Toxin-A
A small number of patients may develop flu-like symptoms. Excessive weakness of the injected muscle can occur, especially when there is underlying weakness masked by the spasticity. This may be a problem in small muscles, such as forearm, where the Botulinum toxin – A may spread to the adjacent muscles. The weakness will reverse completely over time as the body naturally removes the toxin and spasticity returns. Most episodes of excessive local weakness last less than six weeks.

Rarely a child may experience generalised weakness due to a more systemic effect. This is rare and transient. Incontinence with loss of bowel and bladder control can occur after injection into the upper leg muscles. This may manifest as frequency and soiling with onset about a week after injection. The other side effects include dry mouth and pain on injection.

Current Evidence and Recommendation
The assessment of children with cerebral palsy and evaluation of outcomes following injection of Botulinum toxin-A are complex. The involvement of a multidisciplinary team is recommended. The recent international consensus concludes that injection of botulinum toxin-A in children with cerebral palsy is generally safe although systemic adverse events may occur, especially in children with more physical limitation (GMFCS V). The recommended dosage is intermediate between previous consensus statements. The committee further concludes that injection of Botulinum toxin-A is effective in the management of lower limb spasticity in children with cerebral palsy, and when combined with physiotherapy and the use of orthoses, these intervention may improve gait and goal attainment. It is a grade A evidence for the use of Botulinum toxin to reach individualised therapeutic goals for paediatric upper limb hypertonia, grade B (probably effective) for tone reduction following Botulinum toxin injections and grade U evidence (inconclusive) for improvement in upper limb activity and function. Botulinum injections are generally found to be safe and well tolerated with the most common side effect identified as a transient decrease in grip strength.

How Often should We Inject?
About 5-10% of patients on repeated botulinum toxin injections develop clinical resistance. This is likely due to the development of antibodies to the toxin. Our policy is to give injection at least three months between injections to minimise this possibility.

Muscle Localisation for Botulinum Toxin-A Injection
It is demonstrated that palpation is inadequate for muscle identification, especially in the forearm or deep lower limb muscles. Topical analgesia (EMLA cream) with sedation (e.g. Midazolam) is a method to decrease the pain of the procedure.

Electromyography (EMG) was used to control injections
in the upper limb in children with cerebral palsy. However, children with cerebral palsy have a limited ability to co-operate and perform selective movements. The acoustic signal is reduced in sedated children and the painful procedure usually requires several attempts until correct positioning of the needle is achieved. EMG can be useful in children with dystonia to identify muscles that are responsible for dystonic posture or movement.

Electrical stimulation is independent of selective voluntary motor function, and the patient’s ability to co-operate is unaffected by sedation. Therefore it offers better accuracy and practicality in children with cerebral palsy than EMG. The procedure can be time-consuming and painful and is therefore usually performed in combination with sedation.

Ultrasound allows quick visual identification of the target muscle, differentiation from adjoining structures, exact localisation of the needle tip in the desired position of the muscle and real time adjustment of injected volume and number of injection sites according to individual muscle anatomy.

Conclusions

Cerebral palsy is a broad term describing a group of heterogeneous disorders of movement and posture. Although it is an old terminology, it encompasses a group of patients with similar clinical problems, prognosis and treatment. It allows different medical professionals to communicate. Although the brain insult is static, the pattern and severity of the movement disorder may evolve during childhood. The role of paediatricians is to make an early diagnosis and intervene early. Current evidence has demonstrated that Botulinum toxin-A is an effective way in managing lower limb spasticity. Since patients with cerebral palsy have multi-dimensional problems, an interdisciplinary team has been set up in Tuen Mun Hospital to provide them with a comprehensive and holistic care.

References