

Clinical Practic Guidelines for the Management of Cerebral Palsy

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INTRODUCTION

Cerebral palsy (CP) is known since 1827 following the scientific study by Cazauvielh and the French scientist Delpech. However, it was John Little, an English orthopedic surgeon, who had equinus deformity due to polio, brought this disorder into light. He studied its causes and had related them to parturition; this first report by him was published in an obstetric journal. Interestingly, he was refused TA release surgery for his equinus deformity by German scientist, George Stromwyer, who pioneered it. Later, a French surgeon operated upon him. He got impressed with the results and practiced it in children with cerebral palsy. This was the beginning of his interest in the field of CP. This disease has been named after him as Little's Disease also.

Management of children having cerebral palsy is not an easy task. Because, the condition affects various parts of the brain and produces number of neurological deficits. Although according to the definition cerebral palsy is only a physical disability. However, they can have major associated deficits like poor intelligence and abnormal behavior. Further associated deficits affect special sense organs that can give rise to poor hearing and vision. Surprisingly, the list of deficits does not end here; these children also suffer from speech problem, seizures and so on. Not only this motor and other disorders can produce complications like contracture, dislocation, etc.

What is the solution to this wide spectrum of problems? Is it possible to manage them by just a specialist doctor or a therapist? Can we expect a miracle with some sort of brain tonics? Is surgery a ray of hope for all these problems? Shall we not look into their educational aspect when many of them are trainable, and knowing that Ashtaawakra (according to author he is a cerebral palsy child in Indian mythology) had enlightened us with spiritual thoughts. Is it not our responsibility to share and care for them? Controlled emotion's bridle with perfect intellectual direction and drive is the only path of successful management.

Indeed, it is a challenging disorder because of the following reasons: 1. this disorder can be **prevented or controlled, but** is neglected, 2. the damage occurs to the **most complex functional structure** (or the supreme computer) of the universe, the brain, and 3. Damage to the **Developing brain**: Because of this there are many uncertainties regarding the best possible development of the child. Interestingly, it is difficult to ascertain following a particular therapeutic intervention whether it is a natural development or it is due to the intervention, 4. **Damage to brain affects motor system**: The presence of motor system in animals, especially human beings, compared to plants and non living things have made all the difference in the progress and developments on this planet. The mental faculties, definitely superior to all other animals, could not have made any progress without prehensile (hand skill) and the mobility present in them. This suggests damage to an important system of the brain. Noteworthy, this also manifests in various type (spastic, ataxic, etc.) depending on the site of motor system damage.

DIAGNOSIS

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Cerebral palsy is defined as disorder due to non-progressive damage to developing brain leading to motor abnormality.

Cerebral palsy is a static encephalopathy and occurs due to the damage (insult, injury) to developing (maturing) brain (embryo to 12 years).

Risk factors-It has been considered that there are various risk factors present in mother and father of a child having cerebral palsy for example- Repeated abortions, too long or too frequent menses, malnourishment, drug abuse, etc.

There are some studies that have reflected a defect in the sperm. And this may be responsible for defective maturation of brain.

History of brain damage - This is a very important point in the diagnosis of CP. The damage to brain means damage to motor system which is comprised of many parts i.e., pyramidal, extra pyramidal and cerebellar. They may get damaged in combination giving rise to various types of CP for example spastic, dystonic, ataxic, mixed and so on. It is not only the motor system but also other parts of the brain may get affected. They give rise to various associated disorders like mental sub-normality; hearing, language and visual impairments, etc.

Sometimes, the diagnosis of hereditary disorders may be very essential and all the efforts should be made to rule them out.

All the following points should be included in the diagnosis of CP:

I. Based on the site of **motor system** damage; it is classified into following types:

- Spastic
- Athetoid
- Dystonic
- Ataxic
- Choreoathetoid
- Tremor
- Rigidity
- Hypotonic
- Mixed

II. The child having cerebral palsy should be assessed for the other neurological deficits also; severity of all deficits and presence of complications. These clinical points should also be added in the main diagnosis.

A. Associated disorders:

1. Mental sub-normality
2. Learning disability

3. Poor hearing
4. Poor vision
5. Squint
6. Difficulty in speaking

B. Severity

1. Mild
2. Moderate
3. Severe

C. Complications

1. Contracture
2. Dislocation
3. Demineralization
4. Others

How to Make Early Diagnosis of Cerebral Palsy

A baby who has sustained damage to brain and / or has following clinical picture is likely to manifest full blown picture of cerebral palsy:

A baby who remains dull most of the time and has less movements of limbs and head and / or ; cannot move the limb in a purposeful manner instead it just goes in an in-orderly manner (usually in one particular direction only).

The thumb in palm and / or persistent fisting beyond 2 months of age.

Reduced head circumference

Retarded growth

Delayed social smile

Persistence of primitive (early infancy) reflexes beyond 6 months.

Abnormal turning of neck and head

Asymmetry of body posture, movement and reflexes.

Scientific reasons for the need to have early intervention

A developing brain has lot of plasticity and if stimulated can develop neuronal synaptic proliferation and thereby can lead to development of nervous functions. Therefore, early the better.

It has been noted that if the abnormal brain pathways of the damaged brain are repeatedly stimulated, the channels get regularized, they get fixed and then to make alterations in them is very difficult. It means inviting troubles by getting late.

It is interesting to observe different kinds of development in different kinds of environmental stimulation of

genetically same and similar brain damaged people.

TREATMENT

SYSTEMS OF MANAGEMENT AND FALLACIES: There are many modalities of treatment of cerebral palsy like therapeutic exercises, anti-spasticity drugs, orthopedic interventions, neurosurgical interventions and additional (complimentary) therapies. All of them have somewhere some role. However, each system of medicine or therapy claims that it is best. Certainly each one is good in improving only some specific problem or deficit. This depends on the experience of the clinician to choose correct method for a particular problem keeping in mind over all improvement of the child. But, mostly they are used at a wrong time or for a wrong indication. Not only this, professionals who know one particular modality of treatment prefer to treat with that modality only. Because of this there is a bias and the family gets confused. In nutshell we can say that

The professionals are biased with what they know and

The families are confused as to what is the best for their child.

Each system has some potentiality to improve certain deficits but most of them try to treat each and every problem of these children. Say, a child has physical (A), cognitive (B). Speech (C) problem and seizures (D). Specialist 'X' who is best in the management of complaint 'A' starts managing it. The child shows excellent improvement in 'A' problem (say 100% result). But he hesitates to send him to other specialist and treats 'B', 'C' and 'D' problems also. He fails totally while managing these problems (say 0% in each 'B', 'C' and 'D' complaints). So the child will have over all only 25% $(100+0+0+0/4=25)$ * improvement. If we realize this statistics, a number of problems can be easily solved

MYTHS, FACTS AND PAINS: The next problem in management comes due to myths about cerebral palsy. "Some children develop slowly", "don't worry", etc. words of consolation have made the parents to worship the doctors. The same "God", the Doctor, is dishonored after sometime when the parents see that the child is still not walking.

There are innumerable other myths also about cerebral palsy. This delays the diagnosis and also proper therapy on an improper time. With proper consultation and early diagnosis can we obtain perfect results? No. Because, in India, there is not enough infrastructure and manpower to provide multidisciplinary management at an accessible place. In India, perhaps, there are around 25 lakh cerebral palsy children as compared to only 7 lakh in USA. On an average if one centre caters to 250 children there should have been at least 10,000 rehabilitation centers. Surprisingly, most of the capital cities of the country also do not have such centers. Magnitude of the problem is vast.

CHRONOLOGY OF TREATMENT

In India there is an utter confusion in treatment of cerebral palsy people. It includes both selection of method of treatment as well as application according to the age. Author feels that the following protocol is likely to be most beneficial in children having harmful spasticity, a major impairment:

1. Non-invasive

Physiotherapy, occupational therapy

Use of external appliances

Psychological management, special education, cognitive therapy, behavioral modification, self help skill, guidance etc.; Speech therapy

Vocational guidance.

2. Less invasive

a. Pharmacotherapy:

Brain tonics- for few months to few years following insult to brain,

Medicines- for Spasticity, ataxia, seizures etc., Injections of medications like Baclofen (ITB -Intrathecal Baclofen-Pump). Phenol, Botulinum Toxin.

b. Complimentary therapies

c. Spinal cord stimulation.

3. Invasive:

Surgeries e.g. neurosurgery for Spasticity, movement disorder (dystonia) etc.; orthopedic surgery for contracture and dislocation, etc.

Neurosurgical procedures

SPR (Selective Posterior Rhizotomy)

SMF (Selective Motor Fasciculotomy)

Orthopedic procedures

STR (Soft Tissue Release) surgeries

Tendon transfer

There are a few methods, which may be good for a particular patient from medical point of view. However, they may not be good from their family point of view, because they may not be able to afford. Therefore, social medicine comes into importance while managing the case. Say for example rhizotomy (SPR) and fasciculotomy (SMF) cost around Rs. 10,000 to 20,000 where as botulinum toxin (BT) and ITB cost around Rs. 50,000 to 3,00,000/-. There is recurring expenditure also in the later two methods. However, in some cases these non ablative procedures may be the best suitable procedures from medical point of view. The other methods, which are ablative, may not be best suitable. However, if the family cannot afford then the child cannot be left for non-development and to get into complications of spasticity. A judgment by the physician as to what is best for the family is very much expected and that would only motivate the family to sustain the long-term habilitation of the child.

Dystonia in Cerebral Palsy; Sometimes dystonia are confused as psychiatric disorder. A perfect diagnosis is essential before planning the treatment.

There are many drugs to control dystonia. However, they are not effective in all the cases. Therefore, in selected cases of diffuse variety, especially if associated with diffuse spasticity, ITB can be recommended. Presently, such selected cases have shown some beneficial effects. In our series some functional improvement

was also noticed.

In hemidystonic cases and in a few diffuse cases stereo tactor thalamic-basal ganglionic stimulatory or ablative surgery can be considered. The nano technology in future is likely to be quite promising in this group of cases

In a few segmental and focal dystonic cases botulinum toxin and or selective motor fasciculotomy (SMF) might help. In these cases drugs is less preferred because systemic medication for life long is associated with other drug related complications. The orthopedic surgery is invariably associated with recurrence. The use of BT with recurring expenditure is extremely expensive. Therefore, some help can be extended by SMF. In author's series some control on dystonia could be achieved and in a few cases better use of the hand in bimanual activities was noticed.

In nutshell, treatment of the child includes early diagnosis and intervention. Sometimes diagnosis is uncertain in no risk babies or without the history of brain insult. However, without delay when the child has sustained brain damage or has delayed development, neurotrophic drugs (brain tonics) may be started. Although, fool-proof results on the benefits of these drugs are not known. However, harmful effects have not been seen. But, the parents must realize that medicine is not the ultimate; the definitive treatment is through neurostimulation. In a questionable hope of achieving good results through brain tonics, the neurostimulation and other therapies should not be forgotten.

Both physical and mental exercises (Physiotherapy, occupational therapy, cognitive therapy, special education) are essential during most of the period. Neurostimulation method of the therapy, indeed, stimulates brain and improves neuronal functions of the existing brain cells. Brain has tremendous potential power which can be brought to light by stimulating therapies.

Some children despite all these measures show excessive spasticity or movement disorders, they may benefit with drugs. Children with seizures need anticonvulsants regularly and the dose must be increased with increase in weight.

At some stage the child may not improve further and may have harmful spasticity, ataxia or movement disorders. Such children may improve further with complimentary therapies; and if not then neurosurgical intervention is warranted. However, certain cases especially those who have been over looked or neglected, develop contractures, dislocations, etc. These complications are best treated by orthopedic surgeries. But undoubtedly, during all these days exercises are essential. There is no substitute for mental and physical exercises.

Ultimate goal of the management of cerebral palsy people is to provide them as well as their families a comfortable life and to make them as much productive as possible.

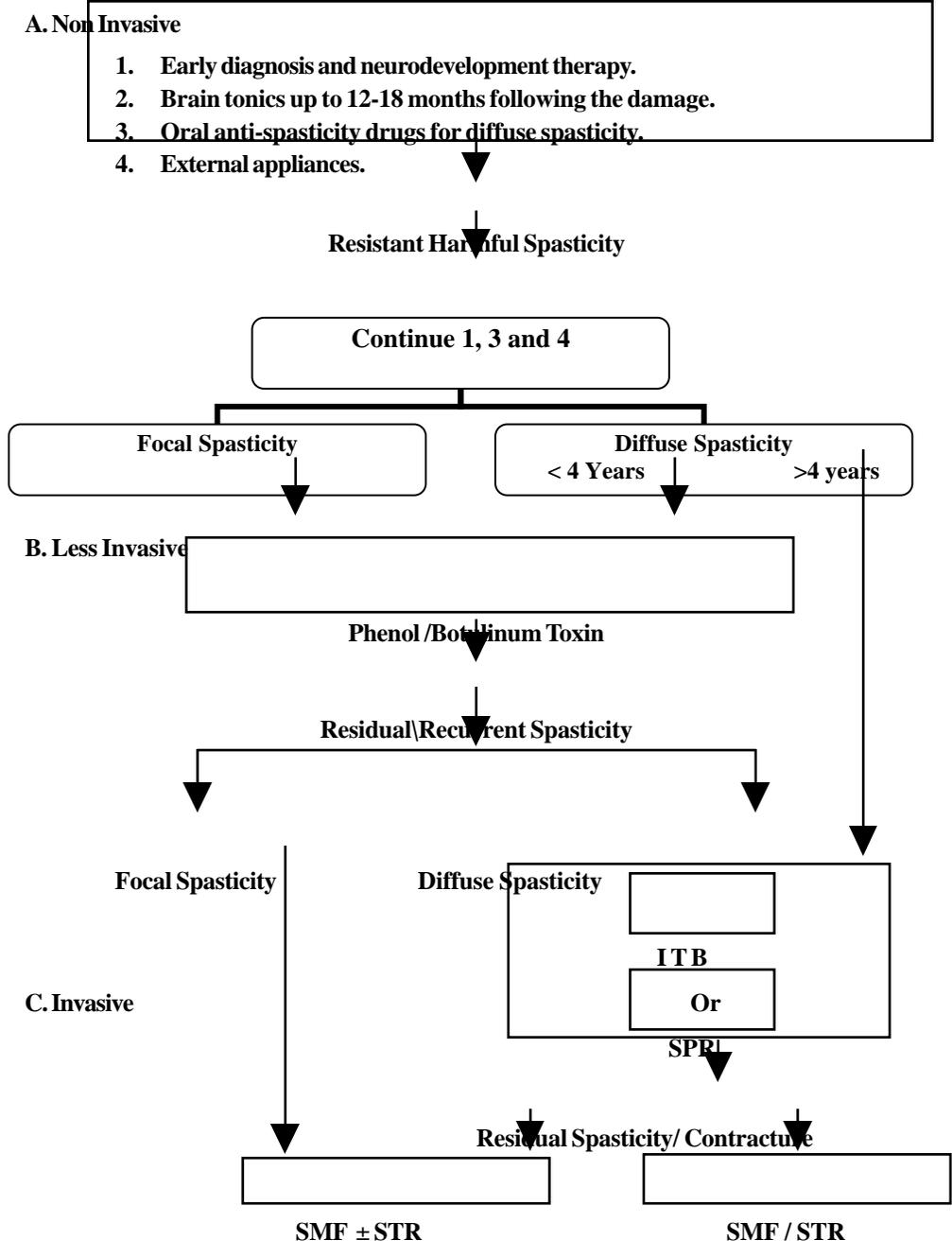
Therefore, perfect vocational training is also necessary.

We need to improve care of cerebral palsy people from a very preliminary stage.

Awareness, early diagnosis and intervention at an unconstrained geographical location are the greatest need of the day for better care of children having cerebral palsy.

To some up it is very important for clinicians to know evaluation of the child with cerebral palsy and to correctly decide the best helpful procedures for that child and the family. It is also essential to acquire knowledge and skill in as many procedures as possible. So that bias for what they know is eliminated. **Dr. A. K. Purohit, Prof. & Head, Department of Neurosurgery, Nizam's Institute of Medical Sciences, Hyderabad, Website:**

Clinical Practice Guidelines for the Management of Spastic CP
I. MOTOR IMPAIRMENT E.G SPASTICITY



II. Associated disorders need treatment at the same time when spasticity treatment is in progress.

III. Complications like contracture is treated following the reduction in spasticity only.

