CEREBRAL PALSY (CP) - LITTLE’S DISEASE

Introduction :-
- It is the most common form of chronic motor disability.
- It is not inherited but some have genetic background .
- Prevalence is 2/1000 life births .
- It is a consequence of damage to the developing brain can result from different causes .

Definition :-
It is a dynamic disorder characterized by presence of all these 4 criteria :

1. The pathological process exclusively affect the brain , not the other part of the C.N.S..
2. It affects motor and posture .
3. The lesion in the brain is static(non progressive and non curable for ever) , but peripheral clinical manifestations may change by time .
4. Should occur during development of the brain (fetal life , infancy , early childhood ) .

Etiology :-
80% of the causes are antinatal, 10%during birth. Only 10% due to post-natal problems.

Prenatal :
1. Intra uterine infection (maternal) .
3. Anoxic .
4. Genetic (syndromes) .

Perinatal :
1. Perinatal stroke .
2. Intrapartum asphyxia .
3. Prematurity .
4. L.B.W. .

Postnatal :
1. Kernicterus (bilirubin enciphalopathy) .
2. Trauma (birth injury , head injury ) .
3. Infection of the brain & menings .
4. C.V.A. .
5. Brain hypoxia .
6. Hydrocephalus .

Classification :-
Physiological :
1. Spastic c.p. (most common) 70% of cases .
2. Athetoid 10-20% .
3. Ataxic 5-10% .
4. Atonic (rare) .
5. Mixed type , e.g; athetoid with spastic .
Topographic classification:

1. **Monoplegic**: only one limb affected.
2. **Hemiplegia**: unilateral upper L. & lower L. affected
   - but upper > lower.
3. **Paraplegia**: both L.L. affected.
4. **Diplegia**: all 4 limbs affected but L.L. > U.L.
5. **Quadriplegia**: 4 limbs to the same extend.
6. **Triplegia**: 3 limbs affected 1 spared.
7. **Doublehemiplegia**: 4 limbs ,but U.L > L.L.

Etiologic:

1. Prenatal (congenital c.p.) when the event occurred during intra uterine life. 80%
2. Natal.(during birth process). 10%
3. Postnatal (acquired c.p.) the event occurred after birth. 10%.

Functional:

1. **Class 1**: no limitation of activity.
2. **Class 2**: slight to moderate limitation.
3. **Class 3**: severe limitation (bed ridden or wheelchair).
4. **Class 4**: no useful physical activity (should be fed and cleaned).

According to the site of the lesion:

1. **Pyramidal tract**: mono , hemi , di , paraplegias occur when the lesion is there.
2. **Extra pyramidal**: spastic, athetoid types.
3. **Cerebellar type**: ataxic c.p. usually anoxic event happened.

Clinical features:-

- Usually most of them they have delayed developmental milestones.
- Absence or persistence of neonatal primitive reflexes are alarming signs, e.g; persistent morrow reflex beyond 6 months.
- Signs of upper motor neuron in the affected limb.
- Other clinical features according to the type of c.ps appear in this way:–
  - All spastic C.Ps are about 60% of cases.
  - Most of them they have no asphyxia, but C.V.A.

Spastic hemiplegic c.p.:

1. Decrease active movement in the affected side.
2. Hand preference at an earlier age(< 1 year old).
5. Equinovarus deformity of foot.

Spastic diplegic c.p.:

1. In infancy they have scissoring of the legs on holding.
2. Commando crawl during crawling.
3. Adduction spasm (causing difficulty in diappering).
5. Few of them have M.R, 25% epilepsy.
Spastic quadriplegic c.p:
1. Most severe form.
2. Most of them have M.R., epilepsy.
3. Some have speech, visual problems.
4. May get pseudobulbar palsy which cause difficulty in swallowing and aspiration pneumonia.

Dyskinetic C.P.(commonly due to kernicterus): Usually the lesion in is in extrapyramedal tract.

Athetoid C.P., Choriform, Dystonic C.P.:
1. They have uncontrolled slow writhing movement of distal part of U.L.
2. Usually they have hypotonia.
3. Head lag is the early manifestation.
4. Feeding difficulty.
5. Epilepsy uncommon, no M.R.

Atonic c.p.:
1. There is hypotonia but brisk reflexes with positive babaniski sign.
2. Present as floppy infant (both ataxic & athetoid type may present in this way).
3. Should be differentiated from myopathies and muscle dystrophies by brisk reflexes.

Ataxic c.p.:
1. There is hypotonia, ataxic gait.
2. Cerebellar signs are positive.
3. May coexist with spastic L.L. called ataxic diplegia.

Diagnosis:-
- Mainly clinical.
- Progressive disease of the brain should be excluded.
- E.E.G. when there is seizure to identify the type.
- Audiometry for hearing assessments.
- Ophthalmological exam for eye problems.
- C.T., M.R.I. not routinely done unless you search for detail of the cause and pathology.

Treatment:-
The aims:
1. Improve function.
2. Prevent deformity.
3. Encourage independence.
- It is team working between pediatrician, physiotherapist, psychotherapist, orthopedician, speech, social occupation therapist.
- Physiotherapy is the main line.
- Pharmacotherapy: drugs used for spasticity like baclofen, benzodiaxepins, botulinum toxin.
- For epilepsy: anticolvulsants.
- Surgery: orthopedic for deformities and contractures.
Complication :-
1. Mental retardation .
2. Epilepsy .
3. Hearing and vision problems .
4. Ptyalism .
5. F.T.T. .
6. Recurrent infections .
7. Contractures .

Prevention :-
- Vaccines and drugs for treatable STORCH .
- Improve safe delivery .
- Prevent kernicterus .
- Proper treatment of C.N.S. infection .
- Prental counseling .

Mental Retardation (intellectual impairment)

It is a significant sub-average intellectual function impairment existing concurrently with deficits in adoptive behavior (communication, self-care, and self-direction). Manifested during developmental period. Incidence about 2% of population.

Classification

- Mild: I.Q 55-70 about 80% of retarded children when become adults they can marry, live independently, be employed and have functional reading and writing. They have deficits in judgment.
- Moderate: I.Q 40-55 about 12%. They require continuous supervision and economic support. They are capable in self-care and employment in sheltered setting.
- Severe: I.Q 25-40 about 7% they are totally economically dependent and needs close supervision. They may acquire language and can be trained in elementary self-care skills.
- Profound: I.Q <25 about 1%. They have limited communication and self-care. They require highly structured environment with continuous supervision.

Etiology

- Mild M.R.: more prevalent in lower socioeconomic groups & rarely explained by a biologic cause.
- Moderate & Severe: more distributed among all socioeconomic classes and more frequently tend to have a biological cause.

Conditions associated with MR

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<th>Type of condition</th>
<th>Example</th>
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<td>Malformation</td>
<td>Encephaloc.</td>
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<td>Prenatal</td>
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<td>Metabolic, toxin</td>
<td>P.K.U., I.e. poi.</td>
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Assessment

- History, F.H., pedigree, prenatal & perinatal H.
- Exam: H.C., dysmorphic feature, anthropometry.
- Developmental history.