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Brain Damage and Long-Lasting Sequels in Childhood: What Does Cerebral Palsy (CP) Mean at the Beginning of the XXI Century?

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1. Introduction

1.1 A great question: The patient and a “legal” definition of CP

In an article from DMCN¹, a question is discussed by many authors, with the aim to consider carefully, CP syndrome versus disease, that is to say a human being suffering from his birth on from a lot of sequels subsequent to brain damage.

The common CP factor consists in permanent posture and movement disorders, with or without mental difficulties, leading to the more patent functional handicap, in standing, sitting, walking, and stiff or impaired upper limb movements. Nowadays, it seems no more adequate to sum up the CP patients by their motor troubles but it is necessary to think over the other factors of limitation to integrate the normal social mainstream at any age, from perception, behaviour, cognition, communication troubles and epilepsy seizures.

CP, an umbrella term? So, might it be really necessary to extend the historical and usual definition of the CP syndrome admitted as sequels of non progressive cerebral damage, whilst may occur during the development of the foetus' brain or of the new born baby's one. From an epidemiological and scientific point of view, it does not seem relevant to include patients suffering from similar neurological sequels but after cerebral tumours or traumatism, brain infections, epileptic attacks, genetic (such as Lesch-Nyhan syndrome, glutaric aciduria disease) and degenerative brain diseases.

In fact, in a medical practise, so called CP patients from different aetiological causes are attended to, cured, treated and followed up in same clinics or medical centres, often with similar rehab techniques, similar drugs, similar surgical decisions on musculoskeletal problems, and secondary orthopedic ones; there, the children's various learning difficulties are analysed to support them in their scholarship. Here, too, the parents may find a necessary help and support to cope with the daily life constraints and talk about their concern in the future with caregivers, social workers and other parents.

1.2 Brain damage, mental capacities and development

Physical (mechanical) limitation and mental development may be different, it depends on the aetiology of the disease, on the type and extent of the cerebral lesions. The independence between poor body state and intellectual power has been pointed out by G. Tardieu who in 1952 described the IMC, (standing for *Infirmitté Motrice Cérébrale* =IMC) as a little part (1/3) out of the global term of CP, known at that time as infantile encephalopathies. The most mental impaired patients were not to be neglected or to be let aside. Tardieu wished to adopt a basic experimental attitude to check his hypothesis. How strong is the voluntary and conscious patient's participation during rehab sessions to recover part of his motor abilities? The subject's role by his interaction is obvious to understand: rehab techniques and relationship with a caregiver allow a better learning and memorizing.

The hypothesis was not checked, now it is known that functional improvement is not completely linked with a mental potential : a lot of CP children with poor mental means are able to walk easily enough -as they can - sometimes by odd pattern even without any rehab session! (see further innate circuitry).

Another important aim, in the nineteen fifties, was a social necessity, to underline and prove that IMC with good mental capacities were eligible to go to normal public schools. In France it was very difficult, even impossible, to go to school with a motor impairment from cerebral palsy*. It may seem stunning to emphasize that mental development is possible without a necessary sensori-motor experience, but that is so! The sole mental representation of action (as described by Piaget), the access to symbolism and abstraction do not come from manipulation but from a process of perception and cognition work. Some athetotic patients (especially kernicterus victims) without any capacity to take and seize an object or to touch it carefully, may reach a high mental level, without any perception trouble. Some of them with limited motor oral means to express themselves, can build up an inner language, learn literacy and are able to typewrite by sophisticated software and to use remote commands. Before electronics performances existed, and sometimes even to day, because it is cheap and easy to make, they are used to show photos, pictures or ideograms from a board or notebook, by visual signs, mimics or simple gesture to pass a message to their parents or friends. *Many CP adults in wheelchairs and using electronic software, best bear witness² to their scholar success or relative failure, to their effort to integrate society or to keep their difficulties in everyday life under control.*

But it is true, risk is high that cerebral damage affects the cerebral networks which participate in the mental development: some recent studies predict about 50% of global cases.

1.3 CP and somatic troubles in child and adult life

Although the cerebral damage itself is not progressive, its clinical expression is "not unchanging" over time. Parents and caregivers know and are afraid of loss of mobility performances, weight of somatic problems, as nutritional failure, feeding difficulties,

* After the 2nd world war many epidemics, all was done for the poliomyelitic children and adults, to let them come back to their previous social status.

sialorrhea, gastro-oesophageal reflux and its complications (insomnia), constipation, respiratory difficulties, sphincterian problems, skin damages. Later, one has to cope with early aging and numerous new health problems, not only psycho-medical ones but also social consequences and lifetime high cost.

One has to try and attenuate them as far as possible!

2. Prevalence

For the last 50 years, when the means of prevention of the neonatal problems and delivery conditions have been organized, the rate of 2 CP children per 1000 children born alive has not changed. In all countries, the most numerous CP are male patients and first born in the family.

The number of premature births is as high as 8%, especially with the babies born at or before 31 weeks of gestational age³.

A lot of recent studies point out the responsibility of prenatal causes of CP. So, up to date research is carried out in the field of neuro-protection of the foetus animal (mice) brain against excito-toxic challenge: the purpose is to apply to the mother and/or new born baby the experiments that have proved beneficial.

3. Etiology

Since 1960, morbidity has been changing. Kernicterus has disappeared, thanks to efficient preventive measures; miserable delivery conditions have been improved by monitoring system, planned caesarean sections; the legal dispositions to protect pregnant women and the quality of neonatal reanimation, in specific units, has played a great role.

Nowadays, more and more patients with CP, around 50% , are ex-premature infants.

15% are term-born but under hypoxic conditions from obstetrical complications or circulatory attacks on the part of the mother or the child or both, before or during delivery.

The hemiplegic palsies represent 20% of the total cases and have often no recognised explanation; the Xray exams often discover impressive cerebral lesion(s), with a real discrepancy according to the state of the baby.

But, it is necessary to know that no radiological cause may be detectable in some infants who have abnormal mental/motor development, it requires a lot of biological complementary exams to determine the pathogenesis of the so-called CP, especially if a hereditary cause is suspected.

4. Early diagnosis of CP is advised

Putting the diagnosis as soon as possible, from 7/9 months after birth, is usual.

For which purpose? From a rehab point of view, the early diagnosis plan is to avoid that the baby has an interval of therapeutic silence, which can be considered as dangerous.

It would allow “to stimulate the brain plasticity”⁴ and so develop the sensori-motor synaptic activity; all these measures would be able to lead the baby to progress and reach a personal autonomy. From a human position, it would also help the parents to deal with their questioning about an unforeseen future.

Here, we shall not explain classical and necessary exam of paediatricians but we shall focus on innovative efforts to understand how the brain damage reveals itself.

We insist that paediatricians have to be aware of not misinterpreting the transient motor abnormalities and CP permanent neurological troubles, in the first year of life.

Many expert clinicians have opened new tracks to find out the first signs of a qualitative motor deficiency, which breaks the natural pattern of the motor development from head - to - toes and allows an early diagnosis.

We cannot really go on speaking about a quantitative psycho-motor delay, because the child is a premature and would need more time to be at a normal level.

The concept of corrected age according to the degree of prematurity must be precisely discussed if applied to ex-premature babies of more than 6/9 months of civil age.

A poor Apgar score at five minutes’ life is not sufficient to predict the future neurological development, it depends on other parameters surrounding prenatal and birth conditions.

The long lasting presence of the archaic reflexes (such as automatic walking) is no longer considered as abnormal. But what is quite different is to observe the presence of asymmetric **neck** reflexes[†], regularly induced by passive head rotation of the patient, and stereotyped , which means a severe brain impairment!

With the young baby under 12/18 months, CP seems like being a disorganisation of the motor development more than a delay in the sequential milestones development. Parents are expecting more and more performances from their baby, who looks “strange”, because he is both, stiff and flaccid, unable to adapt his posture to the mother’s carrying, to shape or control his reactions and actions: to some degree such a CP baby because of his body stiffness may seem to progress better than a normal child, due to his hypertonic up-righting reactions that are considered as very good and advanced performances at such a young age!

Each medical doctor or physiotherapist involved in the diagnosis is used to some different techniques, according to his professional formation, to evaluate how the infant manages its integration between extension and flexion against gravity.

All of them are very careful, knowing a child cannot participate to the assessment because he may be tired from his birth conditions or other somatic reasons or because he is convalescing. At that time, afraid of putting a false positive or negative diagnosis, medical doctors or caregivers are obliged to make a new assessment under better clinical conditions.

[†] different is the asymmetric and changing attitude of a normal baby, known as “fencer attitude”

Is it possible to do a very early diagnosis with a very young and frail premature, because he is at high risk of permanent cerebral damage? Many doctors claim to be cautious and ready to refer to the X ray imaging and other complementary exams to gather items of high risk or to be certain. Indeed, in addition, infant sex and preterm premature rupture of membranes or preterm labour are, also, independent predictors of CP.

Reanimation methods often play a role in restoring the baby's health!

5. The "innate motor circuitry" has modified the knowledge of the infantile motor development

This circuitry differs from the archaic reflexes, which disappear progressively when cerebral high level control becomes predominant. It represents the basis of the involuntary and automatic regulation which is not learnt at the contact with environment, it is done genetically. These pre-programmed reactions, which keep their spatial shape all life long, become more and more efficient, because the muscular bodies have been growing at the same time as baby's will to participate. Some authors speak of maturation of the cerebral system.

Since 1970 approximately, a new conception of the motor development (nativism⁵) has allowed us to insist on the examination of the innate patterns involving structural and global motor organisation (*a celluloid doll body is designed according to the general pattern shaping a baby: the lower limb have semi-flexed knees, the feet are at right angle on the legs, upper limbs with semi-flexed elbows, hands opened; in supine position, in the hand of a girl, the doll "bears its head" aligned with trunk!*)

Briefly, the baby possesses all the natural resources to progress by himself. We know that the basic organisation is obvious in the term born infant and gives him a general motor pattern, which follows a positive evolution during the first year of life. So we can describe a lot of reactions, the head and trunk up righting ones, pull - to - sit, antigravity lateral reactions, ventral suspension, lift manoeuvre, lateral protecting reactions, if someone pushes him on one side...

If the innate circuitry - managing the spontaneous motor expression and automatic reactivity against gravity- failed or is altered at any degree, it means that a more or less important structural injury, is present in the cerebral motor network. This posturo-motor damage is permanent does not depend on intellectual development. The damage cannot deeply be modified by any exercise, passive or active : all life long, because he has lost the automatic quick control, the patient will have to keep or recover his balance under conscious control and voluntary motor command : it represents a relevant clue for care givers to think of very difficult or impossible walking. The patient must progress, while standing by or acting, in using a slow and never perfect correction; if he cannot do it or does not know how or has not a sufficient mental level , he is submitted to the gravity weigh and falls into it.

Moreover, when a baby sinks into gravity, this damage prevents him from acquiring an independence of the axis and limbs. To achieve a rotation of the head, if eye-hand

coordination or spatial orientation is necessary, he cannot pursue a target with his eyes, and all his body is involved in the movement

6. Assessment

Some authors as G Cioni⁶ -following H. Prechtl teaching- are more interested in the attitude patterns of the pre-term infant in the cradle, the variability of the postures, the aspect of the spontaneous movements of the extremities, in time and space. Presence of synchronized and stiff spasms is a sign of neurological disorder when the baby is awakened and does not cry.

Other practitioners are involved in a dynamic examination⁷ exploring the global reactivity against gravity, if the baby general health is sufficient. We can say that during the non – functional period in the baby development, the assessment of the automatic responses opens new tracks towards an early diagnosis. The pathological responses are similar whatever the baby's neurological future will be. The patients who are likely to become hemiplegic can show differences between both sides. Knowing the dramatic developmental changes in the infant's brain, which may question about CP diagnosis, A. Grenier⁸ comes up with an original idea in pointing out the clinical "signs of normality" in the young baby's motor behaviour and communication. His know-how in manipulating the baby to relax him and to lead him to perform some motor coordination, is magical.

Best prediction is achieved through a combination of multiple, complementary tools⁹.

7. Scoring

The well known Gross Motor Function Measure (website: www-fhs.mcmaster.ca/canchild) is one of the method to evaluate globally CP disorder -whatever the neurological trouble is- and to follow up the patient progress; it consists in an examination of 88 items in 5 chapters concerning pro and supine decubitus/ rolling up, all fours crawling / on knees moving, sitting, standing position, walking; the assessment allows to give a score number. This number is reliable if the examiner has been taught by a special instructor, and does not hesitate to ask for a second or third exam, if the results are uncertain.

8. Specific neurological troubles

After the first year of life, as the baby is growing, the classical troubles appear:

CP child seems close to the neurological characteristics of the various cerebral syndromes; they have been classified: hemiplegic form, Little disease (syndrome of premature birth), athetotic syndromes and dyskinetic troubles etc. In the most severe cases, feeding problems, epilepsy, auditive and visual disorders, dysarthria, somatic troubles are very usual. One says generally that CP children escaped psychic disturbances in their childhood, but as teenagers are not unlikely to develop depressive syndromes or trends towards suicide.

8.1 Hemiplegic forms

This unilateral impairment seems to appear classically after a silent interval after birth, but it is not quite true.

Many signs are often pointed out by parents or nurses: they observe that the motor development is a little delayed and that one side of the body does not do what the other does spontaneously, in every day life nursing. for example, to dress the baby. If the practitioner knows how to test the basic organization he can provide items of diagnosis.

From 6 months, it is easier and easier to determine the extent of the hemiplegic palsy because the baby enters the so-called functional period.

The X ray exam confirms the lesions and its extent, usually a cerebral infarctus, inner accident hidden during pregnancy.

In a lot of cases, the motor development is not so much impaired, and lets the parents think of a not so bad general prognosis, by forgetting other factors of cerebral injury. Walking is acquired in the second year of life, but quickly everybody worries about equines posture and other troubles from foot to hip.

Treatment consists in physiotherapy associated to braces, orthosis and botox `infiltrations to reduce factors of stiffness, spasticity included.

We have to confirm presence or not, of lateral homonymous hemianopsia and gnosic troubles in the impaired hand.

Everything has to be checked up as it is important for future autonomy and access to a professional job: side of the impaired hand , remaining capacities to be a grip or a press (30% of the cases) sole use of the forearm (30%). In the last 30% of the cases, the hand is for ever unused or is an ignored tool.

Language most often is saved in right hemiplegic forms, but a following up of the mental capacities is necessary to survey the general development, often not as good as expected to try to prevent school learning difficulties.

Epilepsy is frequent, and often uneasy to be managed by usual drugs.

8.2 Little disease

It is the sequel of 15/20% of the ex -premature infants. We remind that prematurity is defined by a pregnancy of less than 35 gestational age weeks.

The best known disease is the Little diplegia or spastic diplegia, which affects the 2 lower limbs and trunk; apparently the upper limbs are saved , the communication is correct.

In the vertical suspension and lift manoeuver, whereas the normal child spreads his legs, putting his feet at right angle, the pathological pattern is well known: the CP child crosses his legs, and sets his feet in full extension. If the caregiver puts down the child on the ground, the feet remain in equines.

Walking may be acquired later, with a lot of difficulties, such as equines pattern, knee and hip flexed, and hampered by weakness of the trunk. The more difficult walking is, the more frequent orthopedic complications are; all the joint levels of the lower limb may be impaired. The hips are especially affected and need to be followed up regularly, clinically and by X rays.

The rehab management consists in physiotherapy procedures, handling to facilitate corrected positions, use of support positions for sitting, at home or at school.

Soft tissues surgery takes place to maintain a certain quality of the joint motor range and length of the muscles, before a bone surgery becomes necessary. It depends on the functional outcome and patient's way of life.

The complications are more frequent in the severe cases, for example quadriplegia, in which the voluntary command is low or impossible and when the patient is not able to mobilize himself. The participation of mental impairment is to be considered.

Anyway, surgery can modify the local area where it is done, improving flexibility and range of movement but cannot modify the global functional level.

8.3 The associated neuropsychological troubles

The late syndrome of the premature infant has been known, since 1964¹⁰.

It is not a synonymous with motor trouble, because it is a perceptivo-cognitive disorder, subsequent to posterior brain lesions along the visual pathways, handicapping the development of skills that require visuo-mental and planning capacities. Numerous visual troubles are obvious as such squinting, lack of visual fixation and great difficulty in visual pursuit; sometimes, gnosic visual troubles may interfere in the mental representation.

Finally, the child is largely impaired by a great clumsiness, called dyspraxia. Dyspraxia is an execution trouble disrupting a finalized and coordinated gesture, learnt many times by education, it is due to a lack of successive fittings to reach a target; so, in dressing, then using spoon and fork for eating, playing of building up a tower with cubes, making a puzzle, drawing etc.

For example, difficulties in handwriting (dysgraphia) come from a lack of eye-hand coordination and control of the shape of the letters: however, the child is not unable to draw signs provided that they do not correspond to a given shape.

He can symbolize very well about a very poor design.

It is important to highlight that dyspraxia¹¹ should be not diagnosed on a sole symptom but on a net of clues, according to age: parents complaints on poor developmental performances as above mentioned, the results of the psychological tests showing a great discrepancy between language subtests and spatio-temporal ones and the assessment from an advised occupational therapist.

School learnings may be impaired at different levels, except language development which generally is good. As handwriting remains poor and slow, not profitable in time, the use of

an electronic keyboard may be a means to write; but if visual control does not improve enough, the device cannot resolve the top-down problem and entirely compensate the difficulties in performing the task, which still remains slow and faulty. On the other hand, the same cerebral lesions may also affect intellectual capacities in counting and later in mathematics which do not follow an appropriate development.

In case of very great prematurity, under 32 gestational age weeks, the motor disorders are not so frequent but many other neuropsychological troubles may be pointed out involving memory, executive function, attention: as time goes on, secondary and progressive difficulties, related to the strain of school learning and of the necessary high level abilities, heavily bear upon the pragmatic life.

Athetotic syndromes are rare, 7%, including dystonic syndromes.

They are related to hypoxic-ischemia brain lesions on the basal ganglia and thalamus area, in the term-born babies. Kernicterus has disappeared.

The disease looks strange because the patients have mobile spasms, especially on the extremities. All the body, even oral muscles, is often involved, leading to dysarthria, feeding problems and sialorrhea, putting them at a disadvantage.

These patients are not paralyzed but suffering from involuntary movements and spasms, which noise the voluntary gestures forcing them to use strange ones to reach the target. The involuntary movements appear around 18 months; they disappear during rest or sleep time, are enhanced by anxiety or anguish. If the motor areas brain lesions are not extended, they do not provoke neuropsychological disturbances, such as in the late premature syndrome, and allow skills in day life gestures and a challenging behaviour!

But if the lesions are extended further, various CP cases with involuntary movements are associated to many learning disabilities in the field of cognition, communication, and behaviour.

Oral dysfunction leads to many problems, of which underlying components have to be analysed. The lips and tongue adjustments for taking liquids and/or food are impaired, and hamper the stimulation of various swallowing phases. Tongue and face gnosic exam is to be done as soon as possible to complete assessment¹²; sialorrhea and feeding difficulties are responsible for long feeding sessions and cause anxiety and frustration for infant and parents. Then they may lead to a risk of a chronic lack of hydratation, malnutrition, wrong positioning of teeth, and difficult dental hygiene. On the other hand, feeding troubles can induce severe false passages by food choking and determine pulmonary complications. As the CP child does not manage coughing and blowing his nose, he can suffer from ORL chronic infections. Specific cares are to be done to inhibit hyperextension of the body, to facilitate jaw closure and thus feeding sessions and to prevent local complications, sometimes even lethal. In case of the most severe impairment out of any improvement, gastrostomy by tube feeding is discussed.

Speech is impaired or impossible and requires alternative means of communication, from the most simple support to the best sophisticated ones, depending on patient capacities and needs, in order to preserve language development as far as possible.

If the child has access to signing gesture we can consider to learn him Makaton¹³ signs and pictograms to communicate with his peers. The Bliss symbolic resources is a pictographic language for international communication, used for language-related disabilities, not especially for CP impairments.

Classification may be based on topography of the sequels, and one speaks of diplegia, triplegia, quadriplegia ... The principle of classification does not provide clues to treat a complex disease: factorizing must be preferred. The goal is to point out not only obvious deficits but also to detect the remaining, and sometimes hidden, capacities that compose a human being's functioning and behaviour adaptation.

9. Overview

"Parental concern on significant developmental motor delay may well be appropriate while the professional response is dilatory or non-existent" wrote AL Scherzer and I Tscharnuter in 1982¹⁴. This still may be true. But, nowadays, the Xray exams can predict a high risk of persistent abnormalities or assert a fixed motor lesion, early in baby's life. Radiology plays a new role, MRI especially, to appreciate the pathogenesis of the CP. The sophisticated exam may help to put a precise diagnosis but in the great majority of the cases it does not provide information enough to build up a prognosis. But "abnormal" radiological aspects can be misleading: we know new born babies that have been announced to the parents as severely impaired because of abnormal basal ganglia neuro-imaging, whose motor and intellectual development is normal at 4/6 months!

So the clinical exam is not to be neglected¹⁵ and a regular follow up is necessary. Wise and relevant medical attitude is to take time to consider, something such as infant's convalescing time, if all the clues are sufficient for the diagnosis' disclosure.

Anyway we have to remember that there is a CP human being whose progressive needs have to be recognized and taken into account. Parents have to take part in neuro rehabilitation projects: "during the past two decades, awareness of the role of the family in the child's life has increased and the term 'family-centred services' (FCS) ¹⁶ has been introduced to facilitate care for children with special needs and their families". Moreover a new strategy merits further investigation, a therapy approach focusing on changing the task and the environment rather than children's impairments!¹⁷

Anyway, nothing may be done without parents' consent, in the respect of their availability, emotional state, religious faiths. At the end, the financial conditions of the family life must be not forgotten either.

10. Rehabilitation techniques. There is not ONE school of physiotherapy

Conductive Education, Infant Health and Development Program, Infant Behaviour Assessment and Intervention Program represent categories of global approach. Treatment according to Vojta, is envisaged stimulating patterns of active mobilization by nociceptive stimulus, applied by therapist or any voluntary worker without attention to family function.

From the work and publications of Bertha Bobath, other methods are applied, keeping a neuro-developmental action project. In France, many practitioners use the methods taught by Le Métayer¹⁸. He insists on a global motor approach, as in the Bobath method, to teach movements patterns and not a sequence of mosaic of activation of specific muscles.

But, following the G. Tardieu studies on muscle contracture, he advocates to analyze the musculo-skeletal state, fearing that an unsuitable “peripheral tool” interferes with care: the impact of physiotherapy might not be relevant enough.

Indeed, the muscle stiffness must be analyzed and measured: is it an active contracture such as spasticity (defined as velocity-dependant increase in tonic stretch reflex) or other factors included in the upper motor neuron syndrome? Or a passive contracture because of a shortening of the muscle, or a mixed form? We remind that the CP muscle keeps a normal structure and is able to adapt its length to a cast immobilization¹⁹ position, and that muscular shortening means serial reduction of the sarcomeres and modification of the visco-elasticity properties²⁰. If clinical measures are not sufficient, EMG exam can be used to obtain details. Now there is an hype towards a 3D gait analysis to highlight the different intricate causes, but it is reserved to walking patients !

So, according to the muscular properties, it is possible to use passive stretching²¹ by serial casts or other devices to combat soft tissue tightness, (especially the sural triceps) and improve the range of movement, on condition to correct the joint and bones displacements or not to dislocate their structure, in the aim of maintaining the cast. The results are often good during the young age, but do not last, so treatment has to be repeated regularly. After adolescence, passive stretching does not work.

Treatment of the neural and biomechanical components of “increased tone” consists on passive and active movement, positioning in lying, sitting and standing, use of splinting²². Scientific data research on the CP muscle, submitted to strains of the pathological CP motor pattern and its evolution, according to age, is necessary to prevent the muscle from secondary local complications and the limitation of general progress.

In upper limb there are two kinds of treatment: one takes into account the muscular flexibility and the joint/bone orthopedic state, and the second involves the occupational therapist in the management of day life gestures and patient’s motility. The therapists need to communicate between them to let the patient, whatever his age, in the best conditions related to his environment. More and more, the debate is between a child focused-treatment or an environment- focused treatment.

“Everything not useful is finally harmful”.

The goal of botulinum toxin injection is to relax muscles’ stiffness at the point of injection, in reducing local muscular overactivity such as spasticity²³; but it is not a final goal, there must be a purpose behind it; a pragmatic approach is necessary.

So, first of all, how may the patient use or take benefit of removal of spasticity, according to his dysfunction? Spasticity is one of the three components of the upper motor neuron syndrome associated to weakness of the voluntary command and shortening of muscle. Command weakness is often neglected behind the obvious stiffness.

So, we must be sure that this expected result does not provoke too much muscular weakness and finally does not give functional benefit. It is a challenge. It would be interesting to study the other factors which play a role to carry out the rehab project, such as patient's perceptive development, mental capacities, participation, level of interaction with a multidisciplinary team.

Another point: it has been proved, after more than 15 years of practise, that local relaxation improvement thanks to BTX injections has not a long duration, and does not provide functional improvement.

11. Role of medical treatment

For many reasons, drugs are used and very often, to reduce acute or chronic health difficulties. Treatment also concerns management of behaviour troubles, or pain, or epilepsy. On other hand, a lot of oral drugs are required to reduce general hypertony, involuntary movements, as diazepam, dantrolene, baclofen etc. with or without use of BTX. Advantages and side-effects must be analyzed before any new or repeated prescription.

12. Surgery

It would be prudent to keep in mind this phrase "Just one body for one life".

When manual and instrumental physiotherapy means are overpassed, one can think of surgery operations. On soft tissues, it is a tenotomy of various kinds, which can provide flexibility in a sagittal or frontal cross-section. But the younger the patient is, the more transient the benefit is, because of the patient weight and stature growth. In childhood, we can predict a successful result in the short term, *but same causes reproducing same effects*, we can fear the recurrence at long term. The orthopaedic corrections can be discussed, on joint and bone deformities, such as the rotation deformities, hip dysplasia or luxation, fixed flexed knee etc..²⁴

Concerning upper limb surgery, the results are a little disappointing. A reshaped hand and wrist action cannot provide more functional ability than the previous state. BTX injections, associated with surgery to open the palm may be dangerous by reducing fingers strength etc..

Secondary complications on muscle an/or bone are frequent and have to be taken into account and watched over previously, as early as possible.

If hypertonicity is not reduced by previous treatment, neuro- surgical treatments such as posterior radicotomy (or dorsal rhizotomy) or baclofen pump to interrupt the pathways leading factors of hypertonicity can be proposed.

Briefly, any surgical option can provide local or peripheral improvements, but cannot change the functional level; CP patients, with minor or moderate impairments, who spontaneously have acquired aided or non-aided walking and achieved a certain independence, may have a general benefit! The same remark concerns the upper limb operations.

12.1 Overview on outcome

In a remarkable review²⁵, the authors report that there are specific ages to achieve performances: “sitting by 24 m, walking by 72/84 m”. In the CP cerebellar forms, walking with a certain instability may be possible later. The length of time is interesting and shows that a better and earlier rehab intervention cannot deeply change the motor prognosis.

13. Neuro-imaging

Brain abnormalities are present in 70 to 90% of the global cases.

In very preterm infants the standard practise is the ultra sound scanning, with interesting results. MRI[‡] is more sensitive but a difficult exam to perform.

Peri-ventricular-Leukomalacia (PVL) or White matter injury on MRI imaging is conventional.

CP is frequently linked to white matter injury in children born preterm; DTI²⁶ is a powerful technique providing details of White matter microstructure, and reflects disruption of thalamus connections as well as descending pathways.

Findings include decreased grey matter volumes, basal ganglia, thalamus areas, cerebellar abnormalities as well as injury in subplate neurons. With advances in neonatal care the incidence of cystic forms of PVL has fallen whereas the identification of deep white matter injury (non-cystic) has risen.

Dyskinetic²⁷ CP and its subtypes occur mainly in the term born babies, suffering from perinatal insults. In some selected cases, the neuropathological descriptions of status dysmyelinatus, affecting mainly the pallidum in kernicterus, and status marmoratus, referring to the marbled appearance of the striatum and thalamus after hypoxia-ischemia have been described. In great majority, MRI findings are thalamus/basal ganglia lesions, associated to cortical lesions.

14. Alternative devices: Augmentative and alternative means of communication²⁸

It is difficult to give a non-speaking patient, young or not, an alternative means of communication. Parents have built up a specific mode of interaction, assert they understand their child: it is true and not true, it depends on the message. The child 's emotional state, his pleasure or distress, or basic needs may be passed on. But there is high risk not to give a correct meaning to some attitudes or behaviour, associated to a certain vocalizations or noises or handicapped movements. Generally speaking, communication is based on (consistent?) yes/no questioning by the parents who are used to looking for a response from a passive child. But the first communication mode is not sufficient and cannot be adapted to new situations such as going to school and introducing communication with a teacher or friend. To integrate a new system and to implement the communication, one has to work in a multidisciplinary team.

[‡] MRI= magnetic resonance imaging

N Jollieff and H Mc Conachie noted in 1992, at EACD meeting, that four questions can be asked about communication aids: who, what, when, how?

Who. First, a medical exam is necessary to meet the family and let them speak about the antecedents and the diagnosis disclosure of the severe CP disease; feeding problems, often combined with the physical impairment, and their evolution must be assessed. Moreover, visual and hearing quality must be checked, sometimes by a specialist. Then, a psychologist and speech therapist have to evaluate the early infant-mother relationship, child's pre-requisite skills and attention, his verbal comprehension level, whereas the social worker deals with child's and parents' needs.

What. It concerns the type of aid and the means to handle the apparatus, or to touch a keyboard or to use a remote command. So, the occupational therapist is involved in appreciating the limits of the physical impairment, and advising some recommendations. We have noted in the classification's chapter that the devices are various, simple ones allowing the young patient to show pictures in order to communicate with words written under the picture or symbols, other ones more complicated using a grammatical system to make phrases. Some electronic devices are able to give voice to the patient's grammatical work or telegraphic comment. We have to take into account the weariness of the patient and the speed of his contribution and whether the impact on the environment is successful.

Why. The aim is to transfer current communication mode onto more technical system.

How. In many cases, the organization of funding and training has frequently failed because of a lack of an identified co-ordinator, the recommendations have not been followed up, or the patient has not obtained his device (speech synthesizer for example) as expected. The use of computers with adapted remote commands may offer a new facility to those who are able to learn reading and writing, according to their scholarship levels.

The patient's parents and teachers have to be trained to use any alternative device and facilitate the patient's practise.

It is recommended that the multidisciplinary teams keep in touch with alternative and augmentative communication (AAC) ²⁹ organizations (ISAAC organization, for example) to carry out multicentre research in the field.

15. CP teenager³⁰ and adult life³¹

Cerebral palsy is often seen as a disorder involving children only. But children with CP nearly always grow up to become adults with CP, and with continuing improvements in survival. Deaths in children with CP, never common, have in recent years become very rare, unless the child is very severely and multiple disabled.³²

It has become increasingly important to plan appropriate service provision for such adults.

Critical questions that need answering include to review current understanding of physical processes that may contribute to loss of function and premature aging.

It is very important to try to maintain evidence-based care for adults.

Any young adult CP patient who is living in the society mainstream complains that, after years of regular following up in childhood, he cannot find out medical doctor or experienced physiotherapist, or other caregiver, to deal with his needs.

Experienced team is missing also in private or public night and day institutions which attend to CP adults.

Although the neurological injury is non-progressive, CP adults with the disorder often develop well known musculoskeletal complications, but also joint problems, hip/ knee arthrosis, ankle and toes deformities etc. which may lead to severe pain, chronic fatigue, and a premature decline in mobility and function, as they age.

This advance in research of good health and life time span, however heartening, poses new challenges to the medical community.

16. Social cost

In France, the social security entirely covers the CP infants health expenses and medical follow up, physiotherapy charges, equipment and so on. When a CP child enters a rehab and schooling centre, it is free. The most important lifetime costs³³ are the social care costs during childhood.

To inquire about up dated available family aids, in France, according of patient's age and handicap, French parents or parents living in France may refer to the magazine "Déclic" (<http://www.magazine-declic.com/Magazine/>).

The compensatory allowance concerns handicapped people whose disabilities rate is at least 80% and who need the help of another person for activities of daily living, from 20 to 65 years. Later, the handicapped patients receive a special aid.

In France, many associations are involved in supporting actions towards physically impaired people problems[§]. They have created multidisciplinary outpatients and inpatients centres and have organized professional and permanent courses to ensure an updating formation to the carers³⁴, other foundations³⁵ are involved in research on CP causes and quality of care.

17. Pain, distress and quality of life = QOL

The CP life is full of pain(s) and distress periods. Some seem to be related to their chronic disability and physical handicap, some other to the usual human questioning on wasting one's life.

There is a lot of specific questionnaires to help caregivers to understand (highlight) why and where a child or adult patient suffers. For the patients with severe and/or polyhandicapped³⁶ CP impairments, the San Salvadour scale is very useful. There are also self assessment scales to measure pain intensity. But nothing is better than an examination to discover whether pain is precisely situated or extended or long lasting. We know the risk of

§ APF = national association

misdiagnosis, such as unilateral strong back pains vs. renal colic, repeated claims of fatigue, lack of appetite and general malaise vs. severe constipation (fecaloma) etc..

Pain does not come, or seldom, from the physical impairment (except very severe forms or intense involuntary movements) but more often from an awkwardness in parents or caregivers handling, helping to dress, to feed, or to mobilize! There are pragmatic rules to follow regarding the daily life CP assistance and to check if the patient's equipment and orthoses are appropriate for him.

A part of peripheral pain may be related to the moral weight of the handicap and sometimes it is difficult to separate the respective part between physical and moral causes; the complaint may not correspond to the real neurological deficit.

Some patients expect too much from surgical operations and are deeply disappointed of the result, apparently successful, because they have imagined -or dreamt - being cured and not repaired!

However, patients with deteriorated walking function have greater pain frequency³⁷, pain intensity, the impact of pain on daily activities, and physical fatigue and reduced balance. Thanks to a meticulous examination of the patient (neurological, orthopaedic, and somatic exam) and a questioning in the close environment, we can try to resolve a lot of pain sources and reduce uneasiness.

Even though it is difficult, there is always something to do, by medical drugs, psychological support, changing support position, orthopaedic measures. It depends of age, motor level, and social position and patient's will.

Distress. The repeated question from the patients is, why me? And on the other hand, feeling of guilt of their difference and non-achievement in life aspirations and in social integration are factors of a depressive attitude. It is not infrequent to discover alcoholic excess, tobacco addiction or other display of personal discouragement ...

Nevertheless, some adult patients, thanks to their resilience³⁸ property, manage to live the best life they may, by a "CP way of life"³⁹!

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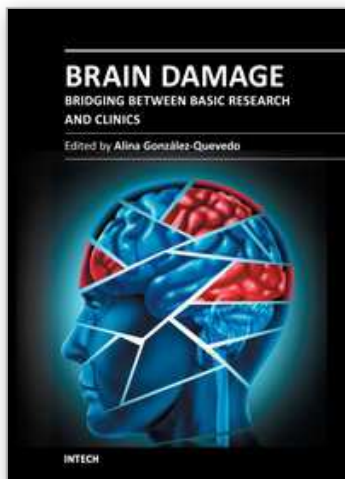
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"Brain Damage - Bridging Between Basic Research and Clinics" represents a collection of papers in an attempt to provide an up-to-date approach to the fascinating topic of brain damage in different pathological situations, combining the authors' personal experiences with current knowledge in this field. In general, the necessary link between basic and clinical neurosciences is highlighted, as it is through this interaction that the theoretical understanding of the pathophysiological mechanisms can be successfully translated into better ways to diagnose, treat and prevent the catastrophic events that occur when the brain suffers from external or internal noxious events. The book spans different aspects of brain injury, starting from damage occurring in the fetal and child brain, followed by different neurodegenerative processes. Attention is also focused on the negative effects of drug addictions and sleep deprivation on the brain, as well as on the early assessment of brain injury for preventive strategies employing sensitive biomarkers.

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