Evaluation of the global development of children with chronic infantile non-progressive encephalopaty

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ABSTRACT

This work researched the mode of acquisition of neuropsychological functions considering the motor, adaptative and linguistic development and the psychosocial behavior by means of specific individualized avaliations. Twenty-seven children carrying non-progressive infantile chronic encephalopathy (CINPE), ages 1-5, diagnosed with cerebral palsy, Down's syndrome, Rubinstein's syndrome and toxoplasmosis, with neuropsychomotor development retardation were surveyed. For the application of the Inventory of Development, a box containing objects varying in shape, size, texture, colors and symbolic value was used. The motor behavior focused on posture, aprehension, movement, general and specific coordination. Adaptation focused on the capacity of adjustment to new activities based on past experiences. Linguistics paid attention to oral communication and/or gesture. Among the psychosocial forms of behavior, aspects such as social adequacy, independence and affection were considered.
Results have shown characteristic patterns for a certain level of maturity. They also have shown that performance is in direct relation to the environment and the possibilities of intervention. Considerable variations from the time diagnosis and in the distinct nature of neurological involvement were observed.

Key Words: encephalopathy, neuropsychology,mental deficiency.

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INTRODUCTION

With the development in neonatalogy the morbility rate is increasing due to a decrease of infant mortality rate (Mancini, 1992) in Brazil and in

the world. In this regard, it is necessary to improve diagnose and stimulation in order to diminish sequels of pre, peri and post natality. It is of utmost importance a stimulating environment to the child development, taking into consideration that sensorial and affective need during the first years of life may induce desaferentiation or functional mental deficiency (Queiróz&Perez, 1992). This pseudo mental deficiency is more severe when associated to specific developmental neurological disorders. Child development occurs in different areas, most of them linked to one another, and are under the influence of organic, maturational and ambiental factors. After injury, it is possible to prevent the compromise of other areas by manipulation of the environment to make it more friendly and stimulating.

An encephalic lesion, in a range of degrees, results in a variety of clinical pictures, chronic or acute or sub-acute depending on the noxic factors acting in the central nervous system.

This condition is called Chronic Infantile non-progressive Encephalophaty and was initially used by Trousseau about a century ago. Chronic means a steady evolution of the clinical picture of the encephalophaty. Non-progressive stands for the non-progressive character of the established anatomopathological lesion. The neuropathological picture is stable but the semiology changes with the child development, as it acquires abilities and progresses in the neuopsichomotor development. This is a characteristic of the Chronic Infantile Non-progressive Encephalopaty (CINPE). Sequel remains and, although child shows its characteristic maturation, in some cases acquisitions may rest stationary in a determined evolutive stage showing increasing discrepancy regarding the chronological age of the child (Diament & Cypel, 1996). The Chronic Infantile Non-progressive Encephalopaties are caused by pre, peri and post natal factors, compromising the Central Nervous System (CNS) in the period of its highest anatomical and functional development.

Among the CINPE we may include Cerebral Palsy (CP), Down Syndrome, Rubinstein Taybi Syndrome, retardment of the Neuropsichomotor development (NPMDR) due to toxoplasmosys, Mental Deficiency (MD) and Learning Impairment due to neurological factors (Barraquer Bordas, 1996).

Cerebral Palsy (CP) is related to sequels of cerebral lesions with persistent, but not invariable, alteration in tonus, posture and movement with onset in the early childhood. The concept of CP is not limited to signals and symptoms due to this primary lesion (non-evolutive) in the brain, but also to signals and symptoms due to the influence of this lesion in the neurological maturation of other encephalic regions (Barraquer Bordas & Verge, 1976). Many epidemiological studies conducted in developed countries have shown CP rates from 1.5 to 5.9/1000 (Bobath & Bobath, 1976). In Brazil, the estimation should be higher since the satisfactory conditions of pre and peri natal assistance are reserved only to a small group of the population. Taking into consideration that the conditions of mother-child care in developing countries are not fully satisfactory, the

infantile mortality rate also reduces the number and register of CP cases. Although the problem is mainly restricted to motor performance, it is possible an association with eye and ear disturbs, sensory alterations, speaking and language disturbs and mental retard (Bobath, 1990).

Down's Syndrome results of chromossomopathy affecting chromossome number 21. From the very begining celular division, the pre natal development is different, leading to a variety of somatic stigmas and a retard in the different steps of the postnatal psychomotor developments. Average incidence of Down's Syndrome is 1/600 births (Casarin, 1997). In general, the clinical picture is typical which makes the diagnosis easy at the moment of birth (Ajuriaguerra, 1976). Although well known, Down's Syndrome is surrounded by prejudice and it is necessary to inform parents and family, teachers and specialists that new researches are pointing to new ways to reduce the levels of deficiency. Neuropsichological investigations aims to define the best approach to the specific moment. Memory, language and attention have a great variability of acquisition and one should always respect the rhythm of each child. Being a non-progressive encephalopathy, it has a spontaneous tendency to improvement. Even without intervention, the child will progress and attain, although slowly, different stages of development, arriving to its limit in a given age, due to the fact that the CNS matures slowly. Thus, the role of the therapeutic team is to accelerate this spontaneous developmental push (Lefévre, 1981).

Rubinstein-Taybi's Syndrome, of unknown etiology, may be characterized by a group of congenital malformations including anomalies of thumbs, halux, cranio facial malformations (hipertelorims, obliquity of eye lids with epicantus, ptosis, nose with wide and flat base, malformed ears with low implantation) and malformations of sexual organs. The palate is ogival which leads to articulatory problems with communication hazards. There is retard in the growth and development with variations in the intensity of the clinical picture as well as in the grade of mental deficiency from one child to the other (Smith, 1989).

The NPMDR in toxoplasmosis occurs due to the contamination of the fetus by *Toxoplasma gondii*, via placenta, with preferential damage of the CNS leading to severe cases of encephalomyelitis. Domestic animals are the main source of infection. In contaminated pregnant woman the risk for fetal contamination increases as progresses the pregnancy and whereas the incidence of severe forms in the fetus decreases progressively from the first to the third semester. There are association of hydrocephaly, convulsions and intracranial calcification which compromises the mental development. Diagnosis can be made by exams such as hemogram, CT, X-ray and liquor. Toxoplasma can be found in sediments of cephaloraquidiane liquid, urine and slides of the bone marrow.

Late diagnosis of the neurological alterations and a lack of attention to the early learning difficulties, due to lack of information or psychological mechanisms of denial on the part of the family, gives rise to severe and, many times, irreversible damage to the development of abilities of the child and in the construction of a better self-stem.

Will the CINPE children be able to acquire the concepts of association, vocabulary, memory, perception discrimination, time and space orientation and other basic abilities needed to the cognitive development?

The main task of mind, according to Piaget (Casarin, 1997) is to assure the adaptation of the individual. The psychological adaptation differs from the organic adaptation by its versatility and efficiency, once it allows the interposition of space and time to achieve equilibrium (Spitz, 1979). Even in a pre-verbal stage, it is possible to recognize important hints of the infantile development, when the child receives some material to which she/he responds or manipulates. The way he/she behaviors to the constraints and conflicts of choice, makes him/her organize his/her own activity and the surrounding world, by means of serial arrangements to attain equilibrium.

When child, despite demonstrating sufficient resources in his/her neuropsischomotor development, shows an unsatisfactory and demotivating performance, with dependency and unstability during its realization, the environment (represented by family) may induce the search for a better understanding of the problem and stimulates the propulsing forces, internal and external, capable of favoring the psychic needs to achieve equilibrium (Amaral, 1994).

Taking into consideration that this population presents multiple deficiencies caused by various etiologies, as well as the need for constant family involvement in the therapeutic process, this study investigated the neuropsychomotor resources of children with neurological sequel, with retard of the global development, characteristic to Chronic infantile non progressive encephalopathy, identifying the aspects of the environment which could be favorable to the evolutive process of the child. The objective was to subsidize future proposals for modification in the pattern of therapeutic intervention aiming a better quality of life for these children.

METHOD

Subject

The study included 27 children ranging from 1 year and four moths to 5 years and 4 months old, being 12 female and 15 male. All children presented neurological deficiency with chronic infantile non-progressive encephalopaty. Diagnosis were Cerebral palsy (14), Down's syndrome (6), Rubinstein-Taybi´s Syndrome (1) and retard in the neuropsichomotor development – NPMDR – due to toxoplasmosis (6) as shown in Table 1. Out of the total, 5 children went to special schools and none were in regular school.

TABELA 1 - Indentification of children participating in the study according to sex, age, diagnosis, and aplies questio waire.

SUBJECT	SEX	AGE	DIAGNOSIS	QUESTIONAIRE
01	F	2 a 5m	PC	x
02	M	2 a 6m	SD	x
03	M	5 a	PC	x
04	M	2 a 4m	PC	x
05	F	3 a 2m	Tx	x
06	M	3 a 7m	SRT	x
07	F	1 a 11m	Tx	x
08	M	1 a 7m	SD	x
09	M	5 a 4 m	PC	x
10	F	2 a	PC	x
11	M	2 a 2m	PC	x
12	F	2 a 6m	SD	x
13	F	3 a 8m	PC	x
14	M	1 a 6m	Tx	x
15	M	1 a 5m	PC	-
16	M	3 a 1m	Tx	-
17	F	1 a 4m	SD	x
18	M	2 a	PC	x
19	F	1 a 5m	SD	x
20	M	1 a 7m	PC	-
21	M	2 a	PC	x
22	M	2 a 2m	Tx	x
23	F	3 a 4m	PC	x
24	F	2 a 8m	SD	x
25	M	2 a 5m	Tx	-
26	F	4 a	PC	-
27	F	5 a 2m	PC	x

Place

Data were collected in the Center for Health Education (CHE) of the Universidade do Sagrado Coração (Sacred Heart University) in Bauru.

Material

It was used an Inventory for Development adapted from the Gessel Scale for Development (Gesell, 1954, 1974; Gesell & Amatruda, 1987) and a family questionnaire specifically elaborated for this study.

To use the Development Inventory it was prepared a box with scraps containing small objects, rubber toys, bell, dolls, glasses with lids of different sizes and colors, books with thick pages, cup, ring, mirror, plastic cup, toy car, sheeths of paper and thick pencil.

Procedure

Data collection

After selection of the target population, an informed consent was obtained from parents to participate in the research.

The Inventory of Development based in Gessel (Gessel, 1954) was applied to the child involving the areas of motricity, adaptation, language and sociability.

Parents were submitted to the questionnaire with information on the identification of the child, school grade and knowledge of the diagnose. In order to investigate the psychosocial behavior parents were requested to fill out a questionnaire with specific questions as shown in figure 1.

1. When and how was the contact with the medical diagnosis?
2. Why do you believe that your child has a deficiency?
3. Which sort of difficulties did the family face in the beginning of he treatment?
4. How is the interaction of your deficient son in the community?
5. Was there important sacrifices (important modifications) in the parents life due to birth of a deficient child?
6. Professional orientation is frequent or sparse? Which is the frequency?
7. Instructions are given to the child, to the family or both?
8. Presently, which are the needs of your child, physical, educational and psychological?
8. Have the family financial resources for treatment? How is the treatment?
9. Are you aware of other multidisciplinary services? Which?

FIGURE 1 – Questions in the family investigation.

At the end of the evaluation parents were informed about the results and those whose children were not under ambulatory care were referred to specific sectors according to the need of the child, including physiotherapy sector, fonoaudiology, psychology and neurology sectors of the Center for Health Education.

Data analysis

Test consisted in tasks that children should normally perform in ages ranging from one month to 3 years and 6 months, based on motricity, adaptation, language and sociability.

In the motor area were evaluated the body posture, domain, coordination, prehension and balance, comparing his/her acquisition with the expected motor development to his/her age.

In the adaptation area the test consisted in the maintenance of behavior facing specific stimulus, auditive and visual attention, initiative and imitation.

Language involved the child's communication to its surrounding by means of mimic, talk, and imitation of sounds, articulation and linguistic organization.

Sociability was studied by interaction of child with the speaker, his/her relation to objects and movements of intentionality for the expression of his/her adaptive will.

For every age there were tests varying in quantity (the average variation of number of item by age was 4) since, depending on the phase, the acquisition are more or less present, many times showing no difference from one month to the next. Thus, the quantity of tests followed the ages. Table 2 shows the maximum scores for each age.

TABLE 2- Expected scores according to age.

AGE (months)	MOTRICITY	ADAPTATION	LANGUAGE	SOCIAL
1	10	4	3	2
2	14	9	5	4
3	20	12	8	7
4	26	15	10	11
5	31	19	11	13
6	37	22	14	17
7	42	26	15	20
8	46	30	16	23
9	51	35	20	25
10	55	39	21	26
11	57	43	22	27
12	60	46	24	30
13	61	50	25	31
14	63	53	28	34
15	70	58	31	41
18	78	64	36	46
21	83	70	39	50
24	88	76	44	57
30	92	83	48	63
36	97	91	53	69
42	100	94	56	71

The application of tests included the following criteria: all tests for the chronological age of the child were performed, giving a plus if he/she could perform the task and minus in case of failure. Each plus was scored 1 point and each minus was zero. If the child reached 100% in the test for that specific age, than the next higher age test was applied till failure of the test. Thus, it was considered 100% of success for the minor age and also 100% of failure for more advanced ages in each area of the development.

The total points in an area (motor, adaptation, language and sociability) represented a quotient for the partial development (DQ). With the arithmetic average of DQ it was obtained the development age of the child (DA). To verify the general quotient for development (GDQ) and the pertaining significance (Table 3) the development age was divided by the chronological age which result was multiplied by 100.

This scoring procedure followed normative criteria as stated by Gessel (1954). The GDQ was considered as a sketch of the comport-

mental development represented by the organization of the developmental aspects and by the habilitation of conduct in the developmental phases in different ages. It was defined by the average of motor, adaptation, language and social behavior.

For a better understanding of the studied material, data were classified in mild, moderate and severe categories, respectively related to the level of independence, need to some assistance and dependency. The latter means that child must be fully cared for by adults in order to survive; assisted means a condition when the child can perform the task but needs instrumental or human support. Independence means that the child is able to perform a task alone. Table 3 shows the different levels of maturity and motor and functional independence.

TABLE 3- Scores, classification and meaning of the performance inventory.

CLHILD CONDITION		
SCORES	CLASSIFICATION	MEANING
Above 180	Normal	Independent
180 – 110	Mild deficiency	Independent
110 – 75	Mild deficiency	Independent / Supported
75 – 50	Moderate deficiency	Assistido
50 – 25	Moderate deficiency	Assistido / Dependente
25 – 0	Severe deficiency	Dependent

The information obtained in the questionnaire were analyzed qualitatively in order to consider the development of the child with CINPE and the psychosocial relation established in the family context.

RESULTS

Results of the developmental evaluation are presented in TABLE 4.

In the studied group, 74% showed a moderate degree of deficiency regarding motricity, compromising the harmony, strength and precision of movements, calling for a constant support for locomotion which characterized a moderate deficiency. In a more severe degree of motor deficiency, 12% showed a marked dependency. However, even with the alteration due to the pathology, 14% were classified average, with independence of locomotion and readiness of coordination.

Behavior of adaptation refered to the action of adjustment that reflected the ability to start new activities and to take profit of the previous experience. In this aspect, closely related with intelligence, all children showed some degree of compromise: 29% with severe, 55% with moderate and 14% with mild mental deficiency.

TABLE 4 – Results of development inventory in the analysed subjects

SUBJECT	AREAS OF EVALUATION				DQ
	MOTRICITY	ADAPTATION	LANGUAGE	SOCIAL	
1	20	20	12	12	75
2	21	19	12	15	76
3	14	11	08	11	21
4	26	18	12	19	26
5	26	24	17	35	86
6	15	32	17	15	58
7	18	17	14	12	100
8	19	18	13	16	106
9	16	10	14	10	25
10	16	18	08	17	91
11	25	28	29	35	180
12	17	20	20	30	106
13	19	19	09	09	36
14	16	15	14	18	100
15	16	23	11	19	100
16	09	05	05	05	18
17	18	34	19	21	167
18	21	10	04	19	79
19	07	06	04	13	45
20	15	12	10	22	142
21	19	18	04	13	75
22	19	13	07	21	84
23	13	18	06	09	35
24	36	40	32	42	168
25	23	23	15	33	109
26	20	07	11	10	31
27	20	10	18	20	35

Evaluation of language was made by use of some type of communication (oral or gestual) with the environment, explored by the interviewer, which could rise some evidence of understanding of the verbal stimulus. Is this aspect, 48% showed severe deficiency, 40% produced just shouts, uttered with hesitation and vocalizations with *dá* (da), *papá* (daddy) and *não* (no) showing an intentional relation of communication representing a moderate level in the language category. Only 7% showed words with some symbolic function.

Social behavior included reaction facing other persons, independence and/or adaptation to family life and social rules such as greetings and caresses. Most of the children (45%) showed uneasiness towards strange people in opposition to a clear recognition and friendly behavior towards members of the family. However, the grade of social and functional independence was impaired favoring the classification of these cases as severe deficiency. 18% of cases were in the mild category and 37% in the moderate category.

The DQ, represented by behavior in the different ages studied, demonstrated 11% showing a pattern of severe global compromise, 33% moderate, 41% mild and only 14% showing a normal pattern of development. This information shows that, despite diagnosis and its implica-

tion, the level of deficiency of these children corresponds to the expected patterns of answers in their age, according to FIGURE 1. Numbers shown in the legends refer to subjects participating in the study.

Twenty-two families answered the questionnaire of investigation about the psychosocial behavior and integration. The difference between subjects and the families participating was due to the difficulty on the part of some parents to attend the interview session, since not only these parents were responsible for conducting the child to ambulatory therapy.

Although the interpreted data followed a criteria with a characteristic pattern of a determinate degree of maturity, some considerations must be made in order to value the child's own personality, the influences of the environment and the possibilities of the proposed intervention. These variables may have represented a constraint in the quality of the performance since a great part of the studied population showed differentiated social, cultural and economical level. There were also relevant variations in the period of diagnosis and the characteristic of the neurological involvement.

According to the report of the family, in the initial contact with the medical diagnoses concerning the condition of these children, the information was given in a very superficial way such as " your son has a problem" " he has some speaking disability", etc. Only with the daily contact with the deficiency did the family decide to return for better investigation and for a better understanding of the inadequacy of the observed performance of the child, which occurred in an average of five months.

The research has also showed that families had poor information on the diagnosis, taking into consideration beliefs and compensatory fantasies. In other instances, they used medical terms without really knowing their meaning or even revealing total ignorance on the condition.

The main difficulties faced by family, from the moment of the child's birth, were mentioned as conflict among parents, anguish by the awareness that something different was happening, and the search for specialized care.

The relations of child in the community turned more oriented, usually by efforts of the mother or a professional. There were reports of avoidance both by parents and members of the community. The relations ocurred more on a child-family basis.

The special child's condition fostered modifications in the family life: the mother discontinued work or study to fully committ to the child; dissolution of matrimony and even statements such as that the new situation has induced personal growth among family members. However, the most frequent data collected concerned difficulties faced by parents toward expenses in order to minimize the financial demands of a costly treatment.

One of the topics made evident by this study was the present needs of the deficient child, since he/she was already under rehabilitation process in some area such as physiotherapy or fonoaudiology. 40% of parents declared that more physical stimulation was required since children did not walk; 31% reported the need for psychological support due

to conflicts on the undue behavior of the child and the attitudes of mother or other members of the family should adopt, despite the lack of understanding about the anxiety and anguish produced by the limiting condition; 13% refered to the need of specialized education in school; 4% declared that the most important was fonoaudiological care to stimulate oral communication; 9% of interviewed mothers were satisfied with the present care program involving physiotherapy, fonoaudiology, neurology, psychology and special education.

CONCLUSIONS

From the experimental point of view, this study aimed at calling attention to some basic elements for the understanding of the development of children with encephalopaty and its relations to the psychosocial environment and, also, to seek formulas to enhance the resources already available, minimizing the individual needs.

The study showed that the physical area is of utmost importance to these children, since it promotes the independence of locomotion, which was not present before. The study also showed that deficiency promoted emotional and psychological instability of parents and their importance in the rehabilitation process to attain the health development of the affected child and family.

To know the functional potential of the child, what he/she learned in his experience and the level of expression of the knowledge, do not mean a precise analysis of his limits, but is important to the search for the basic understanding of the child on the way of evolution. To learn, he needs not only a compatible mental level but also to have the emotional conditions capable of motivating towards action.

Taking into consideration that the psychological needs of the child with CINPE are the same for other non deficient child, he/she needs to experience love, security, approval and the feeling of belonging to the social group in order he/she can structure his personality and to acquire a positive self-stem.

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