Development and psychokinetic therapy of children suffering from West Syndrome – an overview

Gajewska E[1]*, Mojs E[2], Sobieska M[1], Samborski W[1]

[1] Department of Physiotherapy, Rheumatology and Rehabilitation, Karol Marcinkowski University of Medical Sciences, Poznań, Poland
[2] Organization and Management Unit, Karol Marcinkowski University of Medical Sciences, Poznań, Poland

Abstract

Paper presents reasons leading to the West Syndrome, disturbances occurring in the child's development and possible rehabilitation programmes. Psychological rehabilitation of a child with West Syndrome is multilateral, but its main aim is to improve the quality of life, social adaptation and optimizing of the cognitive functioning. Taking into consideration the psychokinetic retardation occurring in the West Syndrome because of prenatal, perinatal or postnatal disturbances apart from pharmacotherapy an intensive psychokinetic stimulation based on plasticity of the brain should be stressed out. Only the complex, intensive and long lasting rehabilitation of a child may secure to it the best possible development.

Key words: West Syndrome, psychological and physiotherapeutic diagnosis, rehabilitation methods.

Introduction

The presented paper is a result of cooperation between psychologist and physiotherapist, therefore it presents more therapeutic than diagnostic approach and has a form of a review.

The West Syndrome (WS) is one of the catastrophic epileptic syndromes in infancy characterised by triad of infantile spasms, psychomotor deterioration and hypsarhythmic EEG pattern [1,2]. WS is commonly associated with poor long-term prognosis outcome, especially in symptomatic cases, with development of other seizure types, impaired motor, cognitive and psychosocial functioning. The aim of our study was to present the therapeutic methods most commonly used in diagnosing and rehabilitation of a child with WS. Early physiotherapy is necessary in all children with WS, but according to the clinical status and possibilities of both therapist and parents various methods may be used.

* CORRESPONDING AUTHOR:
Department of Physiotherapy, Rheumatology and Rehabilitation
Karol Marcinkowski University of Medical Sciences
ul. 28. Czerwca 1956 roku 135/147
61-545 Poznań, Poland
e-mail: karasinska1@wp.pl (Ewa Gajewska)

Received 11.04.2005 Accepted 08.07.2005

Definition of the West Syndrome

West Syndrome is one of the most severe early childhood epilepsy, in which three major components appear: spasms, the EEG pattern of hypsarhythmia and mental deterioration or retardation. Spasms consist of short bend and straighten up or of mixed bend – straighten up movement of trunk, neck or extremities, they may be accompanied by a short shout. Spasms have tendency to appear in clusters of 20 to 40, sometimes up to 100; they are usually symmetric in typical WS. Interictal and ictal EEG [3] consist of generalized high-voltage slow waves and spikes. The treatment of WS is difficult, because the most conventional antiepileptic drugs are ineffective [1]. According to the classification of the International League Against Epilepsy of 1989, WS is classified as general epilepsies with cryptogenic or symptomatic aetiology [4].

According to Smith and Wallace [1] such seizures begin in 90% during the first year of life, usually between 3rd and 9th month, and the morbidity accounts for 1 for 1 900 to 1 for 3 900 children. WS may also present a hidden form in which prognosis both concerning results of the future treatment and first of all proper psychomotor development is much better than in symptomatic forms. Concerning the outcome in WS, the classification in symptomatic and non-symptomatic WS were proposed [5]. Cerebral malformations and neurocutaneous syndrome mainly tuberous sclerosis are the most common causes of WS [6,7].

Early age of occurrence of bend seizures is usually combined with structural disturbances (e.g. cortical displasia), chromosomal or metabolic anomalies (e.g. leukodystrophy) or perinatal aetiology, e.g. anoxia, [2,8,9]. WS may be a consequence of

perinatal hypoxic encephalopathy in about 15% of symptomatic cases [10]. Rarely metabolic encephalopaties are causes of WS, high proportion of these patients have Menkes disease [11] or NARP mutation [12]. Prenatal infections, especially cytomegaloviral infection can cause WS [13]. Matsuo et al. were able to show that in children who were born with low weight (which often means preterm labour) the frequency of West Syndrome is higher than in children with appropriate birth weight [14].

Prognosis in West Syndrome is rather poor, mainly because of diverse aetiology and possible early appearance of seizures of different morphology [15,16]. Prognosis concerning seizures is not optimistic as in about 60% of children epilepsy never cease and in some the Lennox-Gastaut Syndrome with tonic attacks may develop. Psychomotor retardation and other cognitive deficits occur in 80-85% of children in whom West Syndrome was noticed early, before 2nd year of life [1,8]. In 25% of those children additionally a cerebral palsy is diagnosed [8,9].

Despite low mortality, about 10% of all affected children [8,15] the course and effects of the disease make the realization of the developmental tasks and adaptation to the social environment really difficult. Attacks occur in early childhood when the central nervous system is very susceptible to unfavourable external factors [17].

WS symptoms may appear after several months of apparently proper development of the child and then the deficits will be insignificant, noticeable only in specialized diagnostic procedures, psychological or neurodevelopmental.

In therapy of the West Syndrome pharmacotherapy is used most often. Dosing should be adjusted to the patient to affect least possibly its mood and life course. Due to low efficacy of medication also other forms of treatment are used: surgery, ketogenic diet, vagal nerve stimulation [15,18,19].

Evaluation of patients with WS before complex, intensive and long-lasting rehabilitation

Analysis of the reasons of the bend seizures is important mainly for recognition of mechanisms of their occurrence and may have prognostic value, allowing for settings aims of the neuropsychological therapy and kinetic development with higher probability. Mental handicap and other cognitive deficits are caused by the same reason that has lead to epilepsy, but are also a consequence of the disease itself – progressive damage of neurons. Significant diversification of the aetiology of the West Syndrome and of its clinical image – number of seizures, their form, psychokinetic deficits – forms a basis to distinguish a group of patients with better prognosis, so-called gentle bend syndrome [8]. In this group seizures are not so intensive, they occur in later years of life, psychokinetic development is normal or only slightly impaired.

EEG also shows just minor disturbances or is even described as normal. In this patients' group prognosis is rather good, usually the ceasing of symptoms is combined with normalization of the bioelectric activity of the brain and normal psychokinetic development, and thanks to intensive stimulation and neuropsychological therapy even acceleration of selected functions, e.g. speech may occur.

Aim of the treatment process in WS

The aim of the therapy in this particular and in other groups of patients with epilepsy is not just minimising of seizures, but also optimizing of emotional and social functioning of the child.

The treatment process is long-lasting and proceeds in constantly changing psychophysical conditions, also in altering cognitive conditions and interests. Thus psychological proceedings in the West Syndrome should contain systematic and several times repeated diagnostic procedures of the cognitive, emotional and social fitness.

This diagnosis of the cognitive, emotional and social fitness is particularly important in cases of low efficacy of pharmacotherapy, when new anti-epileptic drugs are introduced, mainly because of their side effects, occurring both during monotherapy and in combined treatment due to drug-drug interactions [20,21].

Psychological diagnosis

Psychological diagnosis, particularly neuropsychological gives opportunity of individualized therapeutic programmes and setting aims [22]. Aims of the diagnostic of the cognitive functions are based on monitoring of the course of the disease. Also localizing diagnostics of the brain damages and hemisphere domination is very important. In the psychological investigation special attention should be paid to intelligence level, as according to it further education and its aims are set.

Short-term rehabilitation planning

Short term rehabilitation planning is also one of the aims of the diagnostic [23-25]. Psychological rehabilitation of a child with West Syndrome is multilateral, but its main aim is to improve the quality of life, social adaptation and optimizing of the cognitive functioning. It may be deducted from the clinical practice that parents in the beginning often set too far going aims, often too ambitious and demand to determine advantages and possible probability of their realization. It is not possible because of the diverse clinical course of epilepsy in the West Syndrome, multitude of mechanisms and factors remaining in close dependency and affecting the development.

Psychological diagnosis allows mainly short-term and direct aims. These are set individually and adjusted both to developmental possibilities (developmental plasticity), social possibilities and to the structural limits. Thus the neuropsychological therapy is a form of dynamic interaction of the above mentioned factors.

Learning processes

Learning processes are the main mechanism the neuropsychological therapy in its behavioural aspect is based on. Also some developmental processes, in which cognitive training is a recapitulation of developmental phases of the cognitive

functions, are taken into consideration. A pedagogical approach with mainly behavioural methods, a neuropsychological approach, basing on reorganisation of the functional system, neurolinguistical trying to amplify potential ability to language communication psychosocial aimed on social adaptation and a complex approach, taking into consideration all the above elements [26]. Such an integrated therapeutic process has a long-term character and demands a close cooperation of neurologists, rehabilitants and psychologists.

Physiotherapy

A main task of kinetic rehabilitation is a removal or possibly maximal diminishment of the movement disturbances, as well as gaining control of the kinetic functions in the further development of a child within its largest biologic and social possibilities. Special attention should be paid to locomotive activities, like crawling, or finally walking, manipulative activities and everyday functions. That is why early beginning of rehabilitation of a child with West Syndrome is so important, along with gaining contact with the child itself and its family. A decision about the beginning of physiotherapeutic rehabilitation and about its shape depends on detailed neurological assessment of the child and its behaviour. The whole therapeutic programme must be prepared and modified in cooperation with neurologist and psychologist, because only after complex activity the best results may be expected. Detailed and reliable developmental diagnostics is needed to start physiotherapy, as the individual training programme should be based on the actual assessment.

Munich Functional Developmental Diagnostics

For the evaluation of the development as a marker of complex neurological functions of a child, some methods of assessment of the spontaneous behaviour are used, among others a Munich Functional Developmental Diagnostics, based of the description of the age of: crawling, sitting, walking, grasping, perception, speech development and social contacts development. This Diagnostics is a result of long-term cooperation between paediatricians and psychologists from Children's Centre in Munich. It encompasses experiences based on examination of healthy babies and children. Also data from results of examination of thousands babies with developmental handicap were collected. Basic statement of this method is that every deviation from accepted time ranges or behavioural forms is regarded as a symptom of psychokinetic development disturbance [27].

Philadelphia Profile of the Development

Another diagnostic method is Philadelphia Profile of the Development, prepared by The Institute for the Achievement of Human Potential (IAHP) as a tool serving to set a diagnosis, to work out a programme and to assess the effects of the therapy. It is a table allowing assessing the development of a child concerning perception (seeing, hearing and sense of touch) and executive functions (kinetic, speech and manual fitness) in seven stages of determined age ranges. These ranges correspond to functional levels in the hierarchic structure of the central nervous system [28]. According to this diagnostic a detailed rehabilitation programme is constructed, in which therapy forms a complex stimulation. It relates not only to kinetic, but also to fitness of eyesight, hearing, sense of touch speech and manual. Programme workout for a child with West Syndrome must be done individually. According to Favata et al. the prognosis is generally poor, but thanks to complex diagnostics even slightest deviations may be recognized and that allows the therapists to start rehabilitation in the most crucial phases of the child's development [29].

Importance of individual treatment programme

According to own observation these children present kinetic retardation, often show hypotonia and low spontaneous activity, what causes later problems with head control, intentional grasp, and further with turning, independent sitting down and sitting. They also have difficulties with (crossed crawling and creeping), which is the most important element in the development of the child as it affects not only the development of both hemispheres, but also grasping, sense of touch and eyesight. The therapeutic choice is so rich nowadays that physicians and therapists often make a mistake of showering parents with various training methods, suggesting large number of means that should theoretically help in regaining fitness. It is definitely wrong way. Thus an individual treatment programme should be constructed for each child, with precise description of short-term aims. Such programme constructing often takes a lot of time. Together with parents the therapist has to analyze which therapy should be used as basic, and which one as additional.

The term programme

It should be noticed that better results are often achieved by smaller amount of exercises but conducted systematically and consequently. The term programme means frequency, intensity and time of a stimulus used to create an opportunity for a child to learn and repeat a skill. As every child is different, every programme is different, too, adjusted to particular, individual needs. The highest priority should be given to the skills a child must possess. Then the time needed for every recommended technique should be set.

Doman's method

Doman's method suggests setting a sequence of exercises conducted during each day. Such setting should help parents to conduct exercises systematically and to plan the whole day for their child. Rehabilitation serves to adjust such exercises that by intensity, repetition and action of a stimulus in particular

time should contribute to the child's development. As Glenn Doman states this influence (encompassing intensity, time and frequency) that should compensate at least in part the neurological deviations should be much stronger than the influence of the normal environment on a healthy child. That is why programmes set according to Doman's method may take up to dozen or so hours per day.

Vojta's method

Another rehabilitation programme may be built up according to Vojta's method [30]. It is a rehabilitation less time consuming that may be applied right after the birth if only any deviations are noticed. Vaclaw Vojta, neurologist of Czech origin introduced a method of early rehabilitation in the years 1959-69 already during the stay on Paediatric Intensive Medicine Ward. This method is widely used in rehabilitation of children with various neurological syndromes, in cases of peripheral nerve system damages, in genetic syndromes, asymmetric position or tension. The rules of kinetic complexes described by Vojta apply not only to kinetics, but also on the whole body, its sensual and psychic sphere. Rehabilitation of a child according to Vojta's method starts with an analysis of position and determining of the proper zones of movement initiation that are then stimulated with stimulus of appropriate strength. A child experiences formerly unknown activities, cooperation of whole groups of muscles. It may feel its body much better and builds up a schema of its own body. Thanks to Vojta's methods a reflective turn and crawling may be triggered of. This method is based on the statement that initializing of one kinetic activity and elimination of all pathologic features allows removal of deviations in the kinetic development [8,31]. Building up a programme based on this method one should remember the most basic rules of rehabilitation: definition of the starting position, applying of the adequate stimuli on the precisely determined zones of movement liberation, definition and description of the kinetic response, recurrent character and individual dosage.

Bobath method

Bobath method is another rehabilitation programme, introduced by Karel and Bertha Bobath. This method is very useful in rehabilitation of children with WS and very popular also in children with other developmental abnormalities. The method is based on plasticity of brain and aim to stimulate the development of the child.

The main rationale of this method is based on the statement that the central nervous system regulates the movements, not the muscles, thus one may achieve best results teaching the movements that were initiated by the child itself. Special attention should also be paid to the development of the sensory system that influences the mobility, taking into consideration all kinds of stimuli like proprioceptive or visual. The development of healthy children is regarded as model pattern to assess abnormalities observed in children with disturbances.

Only a therapist with proper education should build up a programme using Vojta's or Doman-Delacato's method, dosing time of exercises and individually set the kind of stimulation.

Conclusions

Proper postural function of the central nervous system is a condition of normal psychomotoric development. This function encompasses automatic, invariable maintenance of the chosen body position. Such a control of body position is inborn and genetically encoded in every human being like also a normal psychic development [30]. Taking into consideration the psychokinetic retardation occurring in the West Syndrome because of prenatal, perinatal or postnatal disturbances (bacterial or viral infections) apart from pharmacotherapy an intensive psychokinetic stimulation based on plasticity of the brain should be stressed out. Only the complex, intensive and long-lasting rehabilitation of a child may secure to it the best possible development. Basing on the experience of many centres the basic form of work of a psychologist and a therapist is to educate parents to make them well prepared therapists of their own child. They – in home conditions – bear the whole task of complex rehabilitation, formerly built up and learnt, both in the sphere of movement and in psychic development. Only thanks to close cooperation of all specialists and parents children suffering from West Syndrome have a chance for better psychokinetic development, and thus – for independency.

References

1. Smith PEM, Wallace SJ. Padaczka – kliniczny przewodnik. 2003.
2. Guberman A, Bruni J. Essentials of clinical epilepsy. Butterworth, Henemann, 1999.
3. Dulac O. What is West Syndrome? Brain Dev, 2001; 23: 447-52.
4. Commission of Classification and Terminology of the International League Against Epilepsy. Proposal for revised clinical classification of epilepsies and epileptic syndrome. Epilepsia, 1989; 30: 389-99.
5. Osborne JP, Lux A. Towards an international consensus on definitions and standardised outcome measures for therapeutic trials (and epidemiological studies) in West Syndrom. Brain Dev, 2001; 23: 677-82.
6. Ohtahara S, Ohtsuka Y, Yamatogi Y, Oka E, Ypshinaga H, Sato M. Prenatal etiologies of West Syndrome. Epilepsia, 1993; 34: 716-22.
7. Rikkonen R. A long-term follow-up study of 214 children with the syndrome of infantile spasms. Neuropediatrics, 1982; 13: 14-23.
8. Michałowicz R, Jóźwiak S. Zespoły padaczkowe i inne stany padaczkowe w zależności od wieku. In: Michałowicz R, ed. Padaczka i inne stany padaczkowe u dzieci. PZWL, Warszawa, 1992.
9. Watanabe K. West Syndrome: etiological and prognostic aspects. Brain Development, 1998; 20: 1-8.
10. Aicardi J, Chevrie JJ. Les spasms infantiles. Arch Fr Pediatr, 1978; 35: 1015-23.
11. Sztriha L, Janaky M, Kiss J, Buga K. Ele and 99 mTc-HMPAO-SPECT studies in Menkes disease. Brain Dev, 1994; 16: 224-8.
12. Makela-Bengs P, Suomalainen A, Majander A, Rapola J, Kalimo H, Nuutila A, Phihko H. Correlation between the clinical symptoms and the proportion of mitochondrial DNA carrying the 8993 point mutation in the NARP syndrome. Pediatr Res, 1995; 37: 634-9.
13. Rikkonen R. Infantile spasm: infectious disorders. Neuropediatrics, 1993; 24: 274-80.
14. Matsuo A, Matsuzaka T, Tsuru A, Moriuchi H, Nakashita Y, Tanaka C, Tomimasu K. Epidemiological and clinical studies of West syndrome in Nagasaki Prefecture, Japan. Brain Development, 2001; 23(7): 575-9.
15. Appleton R. West syndrome: long term prognosis and social aspects. Brain Development, 2001; 7: 688-91.

16. Glauser TA. Following catastrophic epilepsy patients from childhood to adulthood. Epilepsia, 2004; 45: 23-6.

17. Kułakowska Z. Wczesne uszkodzenie dojrzewającego mózgu. Od neurofizjologii do rehabilitacji. Folium, Lublin, 2003.

18. Wheless JW. Nonpharamacologic treatment of the catastrophic epilepsies in childhood. Epilepsia, 2004; 45: 17-22.

19. Campistol J, Garcia-Cazarola A. West's syndrome. Analysis, aetiological factors and therapeutic options. Rev Neurol, 2003; 37(4): 345-52.

20. Mojs E. Ocena funkcji poznawczych u dzieci i młodzieży z padaczką leczonych lamotryginą lub wigabatryną w systemie mono lub politerapii. Epileptologia, 2001; 2: 143-67.

21. Mojs E. Wybrane zagadnienia rozwoju poznawczego a padaczka i jej leczenie. Epileptologia, 2001; 9: 347-58.

22. Schopler E, Reichler R, Dansing M. Techniki nauczania dla rodziców i profesjonalistów. SPOA, Gdańsk, 1995.

23. Mojs E, Kozak I, Brajer S, Zgorzalewicz M. Rola badania psychologicznego w neurologii dziecięcej. Med Dypl, 2000; special edition.

24. Kozak I, Mojs E, Brajer S, Zgorzalewicz M. Diagnostyka neuropsychologiczna zaburzeń poznawczych u dzieci i młodzieży. Med Dypl, 2001; 10: 13-22.

25. Mojs E, Białkowska E. Zaburzenia funkcji psychicznych w padaczce a możliwości edukacji i rehabilitacji psychologicznej w świetle obowiązującej ustawy oświatowej. Pielęgniarstwo polskie, 2002; 1: 95-100.

26. Ognik P. Terapia neuropsychologiczna w klinice uszkodzeń ośrodkowego układu nerwowego – historia poglądów. In: Waligóra B. ed. Elementy psychologii klinicznej. UAM, Poznań, 1995.

27. Hellbruge T, Lajosi F. Monachijska Funkcjonalna Diagnostyka Rozwojowa. Antykwa, Kraków, 1994.

28. Doman G. Jak postępować z dzieckiem z uszkodzeniem mózgu" Poznań. Protez, 1996.

29. Favata I, Leuzzi V, Curatalo P. Mental outcome. In: West Syndrome: prognostic value of some factors. J Mental Defic Res, 1987; 31: 9-15.

30. Vojta V, Die Posturale Ontogenese als Basis der Entwicklungediagnostik. Kinderarzt, 1989; 5: 669-74.

31. Feldkamp M. Krankengymnastik nach Bobath oder Vojta. Kinderarzt, 1983; 14: 857-60.