STEREOTYPED MOVEMENTS OF HANDICAPPED CHILDREN

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I. INTRODUCTION

Repetition is one of the most important factors in learning. It is not too much to say that learning is achieved to a great extent by repetition. Furthermore, repetitive movements and behavior are the most important factors in psychomotor development in normal children. However, in clinical practice we sometimes encounter children who exhibit non-developmental repetitive movements and behavior. As these non-developmental repetitions impede normal development, we must regard them as abnormal behavior.

In the present study we investigated some of these abnormal stereotyped movements of handicapped children with the hope of promoting the development of these children.

II. DEFINITION, CLASSIFICATION AND ETIOLOGY OF STEREOTYPED MOVEMENTS

Stereotyped movements are non-developmental repetitions of identical and rhythmic pattern movements which have no recognized purpose. These movements are not involuntary such as chorea, tremor and myoclonus. The Handicapped children who carry out stereotyped movements usually appear to be fully conscious of what they are doing. We used the term stereotyped behavior to indicate when a child with stereotyped movements walks up and down the room.

Stereotyped movements can be divided into two categories. In the first category, the child employs parts of the body to carry out movements such as body rocking, body rolling, head banging, putting a finger into the mouth, hand fluttering, rubbing the hands, kicking, head and neck shaking, walking up and down, etc. In the second category, the child uses certain objects to achieve movements such as turning over the pages of a book, tearing up a sheet of paper, tapping something on the floor, splashing water playfully, arranging and playing with objects, etc.

Normal children under three years of age normally show stereotyped movements ; however, abnormal stereotyped movements are often noticed in children who : have resided for a long time in homes ; are mentally retarded ; are autistic ; are deaf ; are blind ; who exhibit rubbing hands syndrome ; and who suffer from neuronal ceroid-lipofuscinosis.

We devised the following six hypotheses concerning the cause of stereotyped movements[1].

1) Sensory disturbance : a blind child stimulates himself by using a small amount of

stimulatiom from the outside of the body.

2) Excessive stimulation : autistic children immerse themselves in their stereotyped movements in an attempt to withdraw from excessive and aggressive stimulation.

3) Insufficient stimulation : stereotyped movements are often observed in infant monkeys that have been isolated from their mother and mentally retarded children who have resided for an extended time in an institution for the mentally retarded.

4) Developmental delay : normal children discontinue stereotyped movements as they grow older. Stereotyped movements are often recognized in cases of severe mental retardation.

5) Focal brain dysfunction : repetitive phenomena and perseveration are sometimes noticed due to frontal or temporal lobe damage.

6) Biochemical change : a high dose of adrenocorticotropic hormone (ACTH) or dopamine can induce stereotyped movements in animals or humans.

III. MENTAL RETARDATION

In severe cases of mental retardation, a high frequency of stereotyped movements is observed. The stereotyped movements of these children are generally simple and more frequent after the disappearance of outside human influences and the frequency of the movements decreases when animate stimuli are introduced (such as calling the children by name or making body contact with them.)[2)]

In cases of severe mental retardation associated with frontal lobe damage, (as revealed by CT scans) children who have cortical damage show a tendency toward a high frequency of stereotyped movements.[3)]

IV. INFANTILE AUTISM

Most of the stereotyped movements of infantile autism are characterized by stereotyped walking associated with hand fluttering, head shaking, palilalia, etc. Moreover, the movements are complicated, strange and frequent in comparison with those of mentally retarded children, and they increase with the introduction of animate stimuli as if the children are rejecting any outside human influence. If attempts are made to interfere with or stop these stereotyped movements, the children become confused or panicky. However, some autistic children temporarily stop the movements and then return to repeat them after the disappearance of the outside influence. Autistic infants often develop the tendency to repeat stereotyped movements using objects, but most autistic children under school age only use their body. Autistic children who have no speech show a marked tendency to repeat stereotyped movements in comparison with those who have speech. The first appearance of stereotyped movements is mainly due to : a) changes in the living environment ; b) imitation of another person's behavior ; c) hyperexcitability ; d) isolation for many hours.[4)]

V. RUBBING HANDS SYNDROME (RETT'S SYNDROME)

In 1966 Rett described a syndrome of "Uber ein Cerebral-atrophisches Syndrom bei Hyperammonämie", observed only in girls, which is characterized by autistic behavior, dementia, apraxia of gait, loss of facial expression and stereotyped use of the hands. The

same author reported in greater detail on 21 cases of the same syndrome in 1977.[5]

In 1978 in Japan, we first reported on three cases of girls who demonstrated progressive psychomotor deterioration characterized by peculiar stereotyped movements and autistic tendencies.[6] All of our cases demonstrated peculiar movements such as rubbing of the hands in front of the chest all day long. These girls were quite indifferent and irresponsive to anything happening around them, and they behaved in an apparently robot like manner. Accordingly, we called this syndrome "Rubbing hands syndrome" (Figure 1). Up to now we have encountered 13 girls who have this syndrome. In this paper we describe the symptoms and signs of our 8 patients in tables 1 and 2.[10]

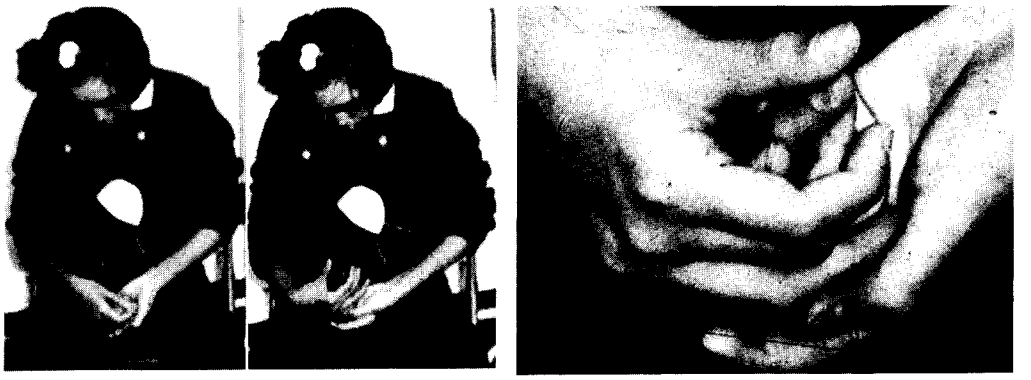

FIGURE 1. A case of rubbing hands syndrome.

TABLE 1 Age at onset of major symptoms in rubbing hands syndrome

Case No. (age)	1 (23y)	2 (22y)	3 (14y)	4 (12y)	5 (8y)	6 (8y)	7 (5y)	8 (4y)
Lack of expression	1y6m	2y	2y6m	11m	1y6m	2y	2y	1y7m
Loss of speech	1y6m	2y	1y6m	1y7m	1y6m	2y	2y	1y3m
Stereotyped movements	4y	5y	2y	1y3m	4y2m	2y	1y7m	1y3m
Gait disturbance	3y m	3y5m	2y6m	1y7m	1y6m	1y	1y7m	2y9m
Attack of epilepsy	3y	2y6m	3y	7y	3y	6m	3y6m	1y7m

The syndrome is marked by rapid deterioration which occurs after a normal psychomotor development at the age of 6 to 24 months. It leads to loss of speech, appearance of stereotyped movements of the hands, autistic tendencies, severe mental retardation and spastic gait, all by the time the child has reached the age of three to five years (Table 1). The other main clinical features are a masked face, holding of the breath, loss of purposeful hand use, epilepsy, bruxomania, hypersalivation, pyramidal tract signs, truncal ataxia, foot grasp reflex, episodic hyperpnea and microcephaly (Table 2).

In Rett's syndrome, the family histories of reported patients revealed no instance of known consanguinity or any abnormal clustering of cases of mental retardation. The majority of cases were born at term following normal pregnancies and uneventful deliveries. The birth weights, birth length and head circumferences were within normal

TABLE 2 Presence of symptoms and signs in rubbing hands syndrome

Case No. (age)	1 (23y)	2 (22y)	3 (14y)	4 (12y)	5 (8y)	6 (8y)	7 (5y)	8 (4y)	Rate
Rubbing hands	+	+	+	+	+	+	+	+	8/8
Holding one's breath	+	+	+	+	+	+	+	+	8/8
Hand grasp disturbance	+	+	+	+	+	+	+	+	8/8
Autism	+	+	+	+	+	+	+	+	8/8
Masked face	+	+	+	+	+	+	+	+	8/8
Loss of speech	+	+	+	+	+	+	+	+	8/8
Epilepsy	+	+	+	+	+	+	+	+	8/8
Jerky movements	+	+	−	+	+	+	+	+	7/8
Spasticity	+	+	+	+	+	+	+	−	7/8
Bruxomania	+	+	−	+	+	+	+	+	7/8
Salivation	+	+	+	+	+	+	+	−	7/8
Hyper-reflexia of DTR*	+	+	+	+	+	+	+	−	7/8
Sensory disturbance	+	+	+	+	+	−	+	−	6/8
Foot grasp reflex	+	+	+	+	+	+	−	−	6/8
Truncal ataxia	−	−	−	−	+	+	+	+	4/8
Tremor	−	+	−	+	+	−	+	−	4/8
Hypotonia	−	−	−	−	+	+	+	+	4/8
Gegenhalten	+	+	+	+	−	−	−	−	4/8
Abdominal distention	−	+	+	+	+	−	−	−	4/8
Babinski's reflex	−	−	+	+	−	−	+	−	3/8
Hyperventilation	−	−	+	+	+	−	−	−	3/8

* DTR=deep tenden reflex

limits. Upon examination, the cerebrospinal fluid had a normal protein content, but the tau fraction, examined in 10 Swedish patients, was markedly decreased or absent in five girls. Blood ammonia concentrations were normal. The electroencephalogram was abnormal in all patients over three years of age. The sleep records contained a large amount of paroxysmal discharges in the form of generalized repetitive slow spike and wave complexes. Motor nerve conduction velocity was within normal limits. The cranial computed tomography was either normal or showed only slight cortical atrophy. The white matter density was normal. Chromosome analysis of the patients revealed no abnormalities.[7]

As the etiology of the syndrome is as yet unknown, we have no effective treatment.

VI. INFANTILE TYPE OF NEURONAL CEROID-LIPOFUSCINOSIS (NCL)

NCL is marked by the accumulation of ceroid and lipofuscin within neurons due to deficiency of a lysosomal enzyme. In 1973 Santavuori reported 15 cases of patients with progressive encepholopathy. This disease had its clinical onset at the age of 8 to 18 months and caused rapid psychomotor deterioration, ataxia, muscular hypotonia, visual failure, microcephaly and myoclonic seizures. All these patients died before reaching six years of age.[8]

Other prominent features were knitting movements. These stereotyped movements were first observed in the hands and forearms when the patients were about two

years of age, but they disappeared after a few months. In Japan, we have not, as yet, found any reports of infantile type NCL. However we can speculate that the knitting movements are similar to those of the rubbing hands syndrome. It is very interesting to note that the knitting movements are specific for brain disease having definite organic lesions.

Diagnostic investigations of NCL include biopsy of peripheral nerves, the appendix and rectum. The electronmicroscopic findings show a curvilinear body, finger print pattern and granular profile within neurons. It is doubtful that the biochemical etiology is attributable to deficiency of peroxidase. There are some reports that treatments of vitamine E or cystine are effective.

VII. ACTH (ADRENOCORTICOTROPIC HORMONE) AND STEREOTYPED MOVEMENTS

We sometimes encounter patients who display stereotyped movements during ACTH therapy against infantile spasms and intractable epilepsies.

One of the authors (Ishikawa, A.) observed three cases of stereotyped movements induced by ACTH. The stereotyped movements appeared at 10 to 14 days after the start of ACTH therapy, and disappeared at five to seven days after the start of the reduction of ACTH administration. I found stereotyped movements such as hand and head shaking, tapping of the lips and putting a finger in the mouth. Moreover, while carrying out the stereotyped movements, the patients displayed a very bad disposition (Figure 2).

FIGURE 2. Stereotyped movements such as brushing her teeth using her two fingers during ACTH therapy.

As the dosage is very high during ACTH therapy, I speculate that administrated ACTH exerts an influence on the neurotransmitters of the brain and induces the stereotyped movements as a natural consequence. In this connection, the removal of the pituitary gland in rats causes learning failure, but supplements of ACTH can help to reverse the failure. A molecular fragment of ACTH without adrenocorticotropic action also improves learning failure in such rats. This result suggests that ACTH acts directly on the mammalian brain and participates in the mechanism of memory and learning. However, as ACTH does not pass easily through the blood-brain barrier into the brain,

very high doses of ACTH (far beyond the normal physiological level) are needed, which possibly produce an undesirable behavioral effect. Direct infusion of ACTH into the rat brain immediately causes characteristic stereotyped movements such as yawning and stretching.[9]

VIII. COMMENT

Herein we described some of the stereotyped movements observed in mental retardation, infantile autism, rubbing hands syndrome, neuronal ceroid-lipofuscinosis, and those caused by high doses of administered ACTH. The symptoms of stereotyped movements are various and their origins are also multifarious in handicapped children. In treating handicapped children with stereotyped movements, it is important to remember that such children have difficulty in establishing close personal relationships. Therefore, we must carefully observe and evaluate the individual characteristics of the stereotyped movements and consider various approaches to assist the child in forming desirable personal relationships.

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