Cognitive Rehabilitation in Pervasive Developmental Disorders

Essay submitted for partial fulfillment of master Degree ***in Neuropsychiatry***

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التأهيل المعرفي في مرضى اضطراب الارتقاء النمائى

بحث مقدم لنيل درجة الماجستير في طب المخ والأعصاب و الطب النفسى

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Summary

Pervasive Developmental Disorders (PDD) are neurodevelopmental disorders characterized by severe and persistent qualitative impairments in three primary areas of functioning: impairments in social reciprocity and engagement; language and communicative skills; and the presence of repetitive non-functional behaviors and stereotyped interests.

These signs start to appear before the age of 3 years old, and in some cases as early as 18 months. The prevalence of autism is about 1-2 per 1,000 people; the prevalence of PDD is about 6 per 1,000, more in males than females with a ratio of almost four to one.

The aim of the work is to review literature discussing pathogenesis, clinical picture, and the principles and targets of different methods of rehabilitation of cognitive dysfunction in autism.

There are several anatomical changes in PDD. There is increase in total brain volume in early childhood, which is heritable by more than 90%. There are focal rather than global pattern of cortical dysmaturation that extends beyond childhood and affects brain regions that are involved in social cognition, language and some aspects of executive function, which is heritable by more than 80%.Also, there is decrease in brain connectivity in frontal and temporal white matter, some evidence in cerebellum, corpus callosum and superior temporal gyrus, decrease in fronto-striatal system, grey matter in left medial temporal lobe, and increase in grey matter in left inferior parietal cortex.

The traditional mentalising theory derives from a symbolic, abstract view of cognition, while the broken mirror account is associated with an embodied approach which

Dedication

To the souls of my dear parents, whom I greatly love and I deeply miss.

First and foremost, I feel always indebted to **God** *the most kind and most merciful who enabled me to accomplish this work.*

I wish to express my greatest gratitude and ultimate thanks to **Prof. Mohammed Hamed Ghanem,** *Chairman of Psychiatric Institute & Professor of Neuropsychiatry Faculty of Medicine - Ain Shams University, for accepting to supervise this work and for his constant help, valuable supervision, and guiding comments. He generously devoted much of his precious time and provided unlimited support, and effort, I sincerely appreciate all the encouragement and support given by him.*

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Marwa E. Khamis

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AAC	Augmentative and Alternative Communication
ABA	Applied Behavior Analytic Interventions
ABC	Aberrant Behavior Checklist
ACC	Anterior Cingulate Cortex
ADHD	Attention deficit hyperactive disorder
ADOS	Autism Diagnostic Observational Schedule
ADOS-G	Autism Diagnostic Observational Schedule Generic
AGTB	Arbeitsgedaechtnis Testbatterie
AIT	Auditory Integration Training
AMIPB	Adult Memory and Information Processing Battery
AMMT	Auditory-Motor Mapping Training
ASD	Autism Spectrum Disorders
ATEC	Autism Treatment Evaluation Checklist
BADS	Behavioral Assessment of Dysexecutive Syndrome
BOLD	Blood-oxygen-level dependence
BORB	Birmingham Object Recognition Battery
CC	Corpus Callosum
CET	Cognitive Estimation Test
COWF	Controlled Oral Word Fluency
DKEFS	Delis Kaplan Executive Function Scale
DTI	Diffuse tensor imaging
EEG	Electroencephalography
EF	Executive Function
EIBI	Early Intensive Behavioral Interventions
FA	Fractional anisotropy
FFA	Fusiform face area
FG	Frontal Gyrus
fMRI	Functional magnetic resonance imaging
HMTW	Hanen's 'More Than Words'
ID	with intellectual disability

List of Abbreviations (Cont.)

IFG	Inferior frontal gyrus
IJA	Initiating Joint Attention
IQ	Intelligent Quotient
ISV	Insufficient Scientific Value
MCST	Modified Card Sorting Test
MD	Mean diffusivity
MEG	Magneto-Encephalography
MIT	melodic intonation therapy
MNS	Mirror Neuron System
MR	Mental Retardation
MRI	Magnetic Resonance Imaging
MRS	Magnetic resonance spectroscopy
N	Number
N/A	Negative association.
NAA	N-acetylaspartate
P	P-value
PDD	Pervasive Developmental Disorders
PECS	Picture Exchange Communication System
PET	Positron emission tomography
PTEN	Phosphatase and tensin homolog
QEEG	quantitative electroencephalograph
rCBF	Cerebral blood flow
RCT	Randomized controlled trial
RCTs	Randomized controlled trials
RJA	Responding to Joint Attention
SD	Standard deviation
SIB	Self-Injurious Behaviors
SIT	Sensory Integration Therapy
SMA	Sequential Meta-Analysis
SMR	Sensory Motor Rhythm
sMRI	Structural Magnetic Resonance Imaging
SS	Savant Syndrome
STG	Superior temporal gyrus
STS	Superior temporal sulcus

List of Abbreviations (Cont.)

TD	Typically developing
TD-controls	Typically developing controls
TEACCH	Treatment and Education of Autistic and Communication Handicapped Children
ToM	Theory of Mind
VE	Virtual environments
VOCAs	voice output communication aids
VOSP	Visual Object Space Perception Battery
VR	Virtual reality
VRT	Virtual reality therapy
WM	Working Memory

Introduction

Not until the middle of the twentieth century was a name for a disorder that affects thousands of children, a disorder that causes disruption in families and unfulfilled lives for many children. In 1943 Dr. Leo Kanner of the Johns Hopkins Hospital studied a group of 11 children and introduced the label early infantile autism into the English language. At the same time a German Scientist, Dr. Hans Asperger, described a milder form of the disorder that became known as Asperger syndrome. Thus these two disorders were described and are today listed in the Diagnostic and Statistical Manual of Mental Disorders as two of the five Pervasive developmental disorders (PDD), more often referred to today as autism spectrum disorders (ASD). All of them are disorders of neural development characterized by varying degrees of impairment in communication skills, social interactions, and restricted, repetitive, and stereotyped patterns of behavior ***(National Institute of Health, 2008).***

These signs start to appear before the age of 3 years old, and in some cases as early as 18 months. Studies suggest that many children eventually may be accurately identified by the age of 1 year or even younger. The appearance of any of the warning signs of ASD is reason to have a child evaluated by a professional specializing in these disorders**.** ASD affects information processing in the brain by altering how nerve cells and their synapses connect and organize; how this occurs is not very well understood till now (***Levy et al, 2009***).

The prevalence of autism is about 1-2 per 1,000 people; the prevalence of ASD is about 6 per 1,000, more in males than females with a ratio of almost four to one. The number of people diagnosed with ASD has increased dramatically since the 1980s. This increase is largely attributable to changes in diagnostic practices, referral patterns, availability of services,

age at diagnosis, and public awareness, though unidentified environmental risk factors cannot be ruled out. A real increase would suggest directing more attention and funding towards changing environmental factors instead of continuing to focus on genetics ***(Szpir, 2006***).

Parents are usually the first to notice unusual behaviors in their child. In some cases, the baby seemed "different" from birth, unresponsive to people or focusing intently on one item for long periods of time. The first signs of an ASD can also appear in children who seem to have been developing normally. When an engaging, babbling toddler becomes suddenly silent, withdrawn, self-abusive, or indifferent to social overtures, something is wrong. Research has shown that parents are usually correct about noticing developmental problems, although they may not realize the specific nature or degree of the problem ***(National Institute of Health, 2008).***

Not many children with ASD live independently after reaching adulthood, though some become successful. An autistic culture has developed, with some individuals seeking a cure and others believing autism should be accepted as a difference and not treated as a disorder (***Silverman, 2008***).

However, no cure is known. Some Children recover occasionally, so that they lose their diagnosis of ASD, this occurs sometimes after intensive treatment. It is not known how often recovery happens; Most children with autism lack social support, meaningful relationships, future employment opportunities or self-determination. Despite tendency of core difficulties to persist, symptoms often become less severe with age ***(Rapin and Tuchman, 2008***).

Abnormal cognitive profile has been observed in people with ASD which demonstrate that ASD is associated with a wide range of cognitive deficits (attention, memory, and

executive functions (EF)) (***García-Villamisar and Hughes, 2007***) .

There are several methods of rehabilitation of cognitive dysfunction as:

Cognitive rehabilitation is done through the application of a lot of procedures in order to enhance development of skills and strategies necessary to overcome cognitive deficits. Computer-based tools can be fruitfully used in the assessment and rehabilitation of cognitive and executive dysfunctions. In particular Virtual Reality could play a key role in the rehabilitation of psychological functions through the creation of synthetic environments where it is possible to carry on tasks very similar to the ones experienced in real contexts ***(Castelnuovo et al., 2003***).

Neurotherapy is also known as Neurofeedback training or brainwave biofeedback. During typical training, one or two sensors are placed on the scalp and one on each ear lobe. Then, high-tech electronic equipment provides real-time (instantaneous) audio and visual feedback about brainwave activity ***(Hammond, 2007***).

Ordinarily, we cannot influence our brainwave patterns because we lack awareness of them. However, when you can see your brainwaves on a computer screen a few thousandths of a second after they occur, it gives you the ability to influence and change them. The mechanism of action is called operant conditioning. We are literally reconditioning and retraining the brain. At first, the changes are short-lived, but the changes gradually become more enduring. With continuing feedback, coaching, and practice, we can usually retain healthier brainwave patterns in most people. Thus, neurofeedback training offers additional opportunities for rehabilitation through directly retraining the brain (***Hammond, 2007***).