Neuropsychiatric Aspects of Salient Child Neuropsychiatric Disorders

Thesis

Submitted for partial fulfillment of the
Master Degree in **Neuropsychiatry**

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1995

Acknowledgement

I would like to express my deep gratitude to my professor *Zeinab Bishry* for her motherly attitude during all my training in the neuropsychiatry department and for her supervision of this work.

My gratitude and thankfulness go also to my professor *Amira Dewedar* for her supervision and meticulous fruitful instructions to me during my residency.

I would like to express my appreciation and thankfulness to *Dr Mohamad Osama Abdul Ghani*, the assistant professor of neuropsychiatry for his supportive attitude and his continuous effort throughout this work.

My deepest gratitude and indebtedness go to our professor *Ahmed Okasha*, the chairman of our department.

Finally, I would like to thank all my professors and colleagues in the neuropsychiatry department of Ain Shams University.

CONTENTS

LIST OF ABBREVIATIONS i

INTRODUCTION 1

CHAPTER ONE: EPILEPSY 5

I. Introduction 5

II. Generalized tonic-clonic seizures in children 8

III. Absence seizures in children 11

IV. Partial seizures in children 14

V. Seizure disorders specific for infants and children 17

VI. Neuroradiological Imaging studies in epileptic children 24

VII. Positron emission tomography studies in children with epilepsy . 26

VIII. Magneto encephalography studies in children with epilepsy 29

IX. Recent genetics of epilepsy 30

X. Cognitive functions in epileptic children 32

CHAPTER TWO: MENTAL RETARDATION

I. Introduction 41

II. Etiology and syndromes of mental retardation 43

III. Electro encephalography and evoked response studies in mental retardation 66

IV. Neuropsychiatric disorders associated with mental retardation .. 72

CHAPTER THREE: CEREBRAL PALSY

I. Introduction 82

II. Etiology of cerebral palsy 86

III. Neuroradiological findings in cerebral palsy 93

IV. Neuropsychiatric disorders associated with cerebral palsy 95

CHAPTER FOUR: AUTISTIC DISORDER

I. Autism and pervasive developmental disorders 98

II. Neurological model of autistic disorder 100

III. Prenatal and perinatal factors in autism 103

IV. Genetic studies in autistic disorder 105

V. Neuroradiological studies in autistic disorder 111

VI. Electroencephalographic studies in autistic disorder 119

VII. Evoked response studies in autistic disorder 124
VIII. Positron emission tomography studies in autistic disorder 127
IX. Neurochemic al studies in autistic disorder 128
X. Neurohistological studies in autistic disorder 130

CHAPTER FIVE: ATTENTION DEFICIT HYPERACTIVITY DISORDER (ADHD)
I. Introduction: Dilemma of minimal brain damage 133
II. Physiology of attention 136
III. Electroencephalographic studies in ADHD 139
IV. Cerebral blood flow studies in ADHD 142
V. Neurochemical studies in ADHD 145
VI. Food studies in ADHD 149

Comment 151
Summary 173
References 175
Arabic Summary

LIST OF ABBREVIATIONS

AAMR	:	American Association of Mental Retardation
ABR	:	Auditory Brainstem Evoked Response
AD	:	Autistic Disorder
ADD	:	Attention Deficit Disorder
ADHD	:	Attention Deficit Hyperactivity Disorder
AEP	:	Auditory Evoked Potential
ALD	:	Autistic-like Disorder
AMLR	:	Auditory Middle Latency Response
ATP	:	Adenosine Tri-Phosphate
BEAM	:	Brain Electrical Activity Mapping
BFNC	:	Benign Familial Neonatal Convulsions
CNS	:	Central Nervous System
CP	:	Cerebral Palsy
CPS	:	Complex Partial Seizure
CT	:	Computerized Tomography
CTX	:	Cerebrotendinous Xanthomatosis
DS	:	Down Syndrome
DS	:	Dissiminated Sclerosis
DSM III R	:	Diagnostic and Statistical Manual of Mental Disorders. Third edition. Revised.
DSM IV Options:		Diagnostic and Statistical Manual of Mental Disorders: Fourth edition options.
EEG	:	Electro-encephalography
EMG	:	Electro-myography
GIT	:	Gastro-intestinal Tract
GTCS	:	Generalized Tonic Clonic Seizures
5-HIAA	:	5-Hydroxy Indole Acetic Acid
HPRT	:	Hypothathine-guanine Phosphoribosyl Transferase
HVA	:	Homo-Vanilic Acid
ICD10	:	International Classification of Diseases. 10th edition
ILAE	:	International League Against Epilepsy
IQ	:	Intelligence Quotient
JME	:	Juvenile Myoclonic Epilepsy
MEG	:	Magentico Encephalography
MHPG	:	Methoxy-Hydroxy-Phenyl-Glycolic acid
MLD	:	Metachromatic-Leukodystrophy
MPSs	:	Muco polysaccharide metabolism disorders
MR	:	Mental Retardation
MRI	:	Magnetic Resonance Imaging
NE	:	Nor epinephrine
PET	:	Positron Emission Tomography
PET-FDG	:	Positron Emission Tomography-Fluoro-deoxy-Glucose
PME	:	Progressive Myoclonic Epilepsy
PKU	:	Phenyl Ketonuria
rCBF	:	regional Cerebral Blood Flow
REM	:	Rapid Eye Movement

SEP	:	Somato-sensory Evoked Potential
SIB	:	Self-Injurious Behavior
SPECT	:	Single Photon Emission Computerized Tomography
SPS	:	Simple Partial Seizures
SSP	:	Subacute Sclerosing Panencephalitis
TCI	:	Transient Cognitive Impairment
VEP	:	Visual Evoked Potential

INTRODUCTION

INTRODUCTION

Neurology and psychiatry were blended as one part of medical science, namely neuropsychiatry in 1800s. However the two sciences were separated and grew increasingly distant after world war II. The neurology became the science of visible organic disease and psychiatry became the science of behavior disorders. In recent decades, the two specialities are again coming together. There is increasing recognition of the biological and neuropathological abnormalities associated with the mental disorders, that is better called now neuropsychiatric disorders. (Solmon and Masdeu, 1989).

Child neuropsychiatry is a subspeciality of neuropsychiatry that manifests clearly this union between neurosciences and behavioral sciences and necessitates the associated efforts of both psychiatrists and neurologists. This subspeciality involves heterogeneous group of disorders in which both psychological and biological factors interacted with each other in their pathogenesis. So it is becoming apparent and accepted by most child neuropsychiatrists that the genetic, chemical, electrophysiological and neuropathological factors contribute to many of the child neuropsychiatric disorders that have been previously thought to be caused by environmental factors alone. On the other side, it is not neither logic nor beneficial to ignore the role of psychological and environmental factors in these disorders. The over simplistic explanations that attribute all symptoms and behavior to a single mechanism whether it is biological or psychological are no longer acceptable. (Shaffer, 1989).

In this work we selected five salient neuropsychiatric disorders in children to review the recent literature in them and illustrate this phenomenon of interplay between biological and psychological factors. These disorders are epilepsy, mental retardation, cerebral palsy, autistic disorder and attention deficit hyperactivity disorder. From our point of view these disorders have the following characteristics in common. These are i) All of them are common disorders (except the autistic disorder).

They are encountered in everyday clinical practice. ii) They present to a child neurologists, child neuropsychiatrists, paediatricians and educational psychologists. iii). Their pathogenesis involve both biological and psychological factors. iv). The theories underlying their etiology are still not settled. v) Management in most of the cases require combined efforts of child neurologists, child psychiatrists, educational psychologists, social workers and sometimes physiotherapists and speech therapists.

As regards epilepsy, it is a disorder of brain function characterized by recurring seizures of various types. It is caused by a wide variety of brain diseases, the final pathway of expression being an intermittent paroxysmal excessive and disorderly discharge of cerebral neurons. (O' Donohoe, 1984). This disorder is one of the most common disorders in neuropsychiatric clinics. The prevalence in United Kingdom and United States of America is around 4 per 1000 (Kangesu, 1984 and Courn et al, 1989). In Egypt, Okasha (1988) estimated it to be four to five times more than in the western countries. i.e. 1.5 to 2 per 100 children.

Epilepsy is not merely a neurological problem. It is commonly associated with behavioral, emotional and cognitive problems. The research in this area is extensive in the last years. (Trimble, 1990, Abdulghani , 1994).

As regards mental retardation, inspite of the general agreement of being a brain disease of heterogeneous etiology, there is still some evidence of a role for psychological factors in its causation. There are evidence from the study of parents of children with mental retardation, adoption studies and twin studies that intelligence is not exclusively a function of heredity or environment. It is clear that environment can either enhance or depress intellectual development. At the same time, the inherited biological factors play their role with the environmental factors to produce the wide variation of intelligence in all people (Baroff 1986).

Second important point in the field of mental retardation that occurred in the last few decades is the shift of the role of neuropsychiatrist. Previously, psychiatrists primarily diagnosed mental retardation and its syndromes. Recent developments in medicine, genetics, biochemistry and behavioral sciences, have made the diagnosis of the medical and neuropsychiatric disorders associated with mental retardation more complex. Neuropsychiatrists now have to recognize a lot of syndromes of mental retardation as well as a lot of associated mental disorders of the same type as encountered in non mentally retarded children except that their clinical manifestations may be modified by their developmental level. (Young et al, 1989).

Cerebral palsy is a disease which does not share the other four selected neuropsychiatric disorders in their interaction between biological and psychological factor in their pathology. However, it is included here because of its high association with epilepsy, mental retardation and other behavioral problems (Eicher and Batshow 1993).

As regards autistic disorder, its recent history goes back to Leo Kanner in 1945 who reported eleven cases of psychoses with onset in the first tow years of life and he labeled it early infantile autism. Psychoanalysts in the 1940s and 1950s were believing of a disturbed infant-mother relationship as a direct cause of autistic disorder. (Young et al, 1989). Nowadays there is a strong shift in this concept. Most neuropsychiatrists believe in the biological basis of austism. There is interplay between genetic, chemical and neuronal structural pathology in autistic disorder. However the exact nature of its neuropathology is still not understood. (Folstein and Piven, 1991).

Attention deficit hyperactivity (ADHD) disorder is a disease of CNS which has a long history of conflict between psychological and biological theories. It had also acquired different names which reflect this conflict starting from minimal brain damage and minimal cerebral dysfunction which reflect the concept of neurological basis.

On the other side, it was labeled hyperkinetic child syndrome and ADHD which reflect the emphasis of concept of behavioral abnormalities.

AIM OF WORK

This work has the following objectives:

i) Reviewing the current knowledge of the etiology and associated pathology of these five disorders with emphasis on the recently described syndromes especially in epilepsy and mental retardation.

ii) Illustrating the interface between neurosciences on one side and the behavioral sciences on the other side in these neuropsychiatric disorders.

It is important to mention here, that our review is not comprehensive about the five disorders. The parts including the epidemiology, clinical picture will be mentioned in brief. Our focus will be on the knowledge that illustrate the areas of interface between biological and behavioral sciences.

CHAPTER ONE : EPILEPSY

EPILEPSY

I INTRODUCTION TO EPILEPSY

DEFINITION

Epilepsy is a disorder of brain function characterized by recurring fits (seizures). It is a symptom of brain malfunction that can be caused by diverse of disease processes, the final common pathway of expression being an intermittent, paroxysmal, excessive and disorderly discharge of cerebral neurons. (O' Donohoe, 1984).

EPIDEMIOLOGY

Reported prevalence rates of epilepsy in children range from 2.5 to 121 per 1000 with most clustering around 4-6 per 1000. (Cowan et al, 1989). Kangesu (1984) reported the prevalence rate of epilepsy in UK. It was 3.4 per 1000 in children aged 5-15 years. Cowan et al (1989) studied the prevalence of epilepsy in children and adolescents in USA. The overall rate was 4.71 per 1000 with the highest rate in children aged 1-4 year. The prevalence rate was slightly higher in boys than girls. The boys were 4.95 per 1000 while girls were 3.29 per 1000 until after the age 14 year when rates are approximately equal. The highest male/female ratio 1.5/1 was observed in infant aged less than 1 year.

Okasha (1988) reported that the prevalence of epilepsy in developing countries is about four to five times the prevalence in developed countries. This may be related to the poor antenatal care, prematurity, birth injuries, malnutrition, multiple infections.

There are some Egyptian studies about the epidemiology of epilepsy in Egypt. Mekky et al (1981) reported a prevalence rate 4.1 per 1000 population. The prevalence was highest in the age group 10-19 years where it reached 7.4 per 1000. In another study El-Afify and Mostafa (1981) on general hospital population, reported prevalence rate of epilepsy 9.87/1000 population. The age specific prevalence was highest in the first and second decades of life when it reached 11.17 per 1000 and 14.4 per 1000 respectively.

CLASSIFICATION OF EPILEPSIES

The International League Against Epilepsy (ILAE) proposed a basic division of epilepsies in 1981, that was refined recently in 1989, to include the numerous, narrowly defined syndromes and several non specific categories into which all generalized and partial seizures should fall. The idiopathic generalized epilepsies are classified based on the age of onset and combination of seizure types. Partial seizures are attributed to restricted sites of the cerebral cortex, predominantly according to their clinical features with supportive evidence from electroencephalography. The principles of classification were established based on the literature, ictal telemetry recordings and the experience of a panel of experts. (Manford et al, 1992).

THE ILAE CLASSIFICATION OF EPILEPTIC SYNDROMES

1. LOCALIZATION-RELATED EPILEPSIES

1.1 Idiopathic (with age related onset)

- ⇒ Benign childhood epilepsy with centrotemporal spikes
- ⇒ Childhood epilepsy with occipital paroxysms
- ⇒ Primary reading epilepsy

1.2 Symptomatic

- ⇒ Chronic progressive epilepsia partials continua of childhood
- ⇒ Syndromes characterized by seizures with specific modes of precipitation
- ⇒ Temporal, frontal, parietal and occipital lobe epilepsies

1.3 Cryptogenic

- ⇒ Temporal, frontal, parietal and occipital lobe epilepsies

2. GENERALIZED EPILEPSIES AND SYNDROMES

2.1 Idiopathic (with age related onset)

- ⇒ Benign myoclonic epilepsy in infancy
- ⇒ Childhood absence epilepsy/juvenile absence epilepsy
- ⇒ Juvenile myoclonic epilepsy
- ⇒ Epilepsy with generalized tonic-clonic seizures on awakening
- ⇒ Syndromes characterized by seizures with specific modes of precipitation
- ⇒ Other idiopathic generalized epilepsies