### INVOLUNTARY MOVEMENTS IN

### CHILDREN

Pathophysiology - Types & Management

**ESSAY** 

Submitted for Partial Fullfillment of the Master Degree in

Neuro Psychiatry

618.92 83 H.A

Presented by

Hanaa Abdel Wahab El Sayed

M. B. B. Ch.

Supervised by

Professor Dr.

Samiha Abdel Moniem

Professor of Neuropsychiatry

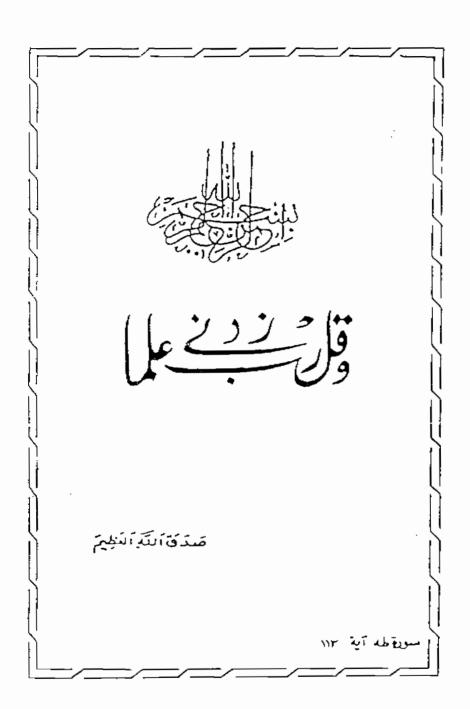
Professor Dr. Zeinab Bishrv

D - 6 - - - - - 6 N - - -

Professor of Neuropsychiatry

Faculty of Medicine Ain Shams University 1986.

1





To My Daughter

## ACKNOWLEDGEMENT

I would like to express my deepest gratitude to Prof. Dr. Mahmoud Mostafa, Prof. & head of neuropsychiatry department, Faculty of medicine, Ain Shams University, for having given me the chance to undertake the subject of this essay.

I would like to thank my eminent Prof. Dr. Samiha Abdel Moniem, Prof. of neuropsychiatry, faculty of medicine, Ain Shams University, who suggested the sbject of this essay. She devoted a lot of her valuable time in helping me with every detail in this work which, without her guidance & help, would never have reached this degree of completion.

I am also greatly in debted to Prof. Dr. Zeinab Bishry, Prof. of neuropsychiatry, Faculty of medicine, Ain Shams University, for all the help, advice & meticulous revision of all the details I received. It was due to her efficient supervision, patience & cooperation, that this essay has materialized.

I would like to take the apportunity to thank my husband Col. Engineer Nashaat Gaafar & my mother from whom I have received great help & encouragement.

I would like also to espress my deep thanks to my father the ex-inspector general of English for his encouragement & for revising the language of this essay.

Finally my deepest thanks are due to all the staff members of Neuropsychiatry department for their helpful intrest & kind support.

Hanaa Abdel Wahab

Central Library - Ain Shams University

# Contents

		۲.
Inti	roduction	1
The	extra pyramidal system	2
Phys	siology of the basal ganglia	12
Neur	robiochemistry of the basal ganglia	18
Path	hophysiology of movement disorders	26
Туре	es of Involuntary Movements .	
1-	Athetosis	36
2-	Chorea	44
3-	Choreoathetosis	58
4-	Ballismus	63
5-	Dystonia	66
6-	Myoclonus	77
7-	Involuntary movements accompanied with kwashiorker.	92
8-	Tremors	96
9-	The Boble head doll syndrome	104
10-	Spasmus nutans	107
11-	Paroxysmal toricollis of infancy	108
12-	Tics	110
13-	Gilles de la Tourette syndrome	115
14-	Juvenile parkinsonism	125
15-	Wilson's disease	129
16-	Mirror movements	134
	- Summary	138
	- Refrences	140
	- Arabic Summary.	140

REVIEW
OF
LITERATURES

### INTRODUCTION

Movement disorders have become a particularly exciting area of clinical neurology because of many recent pharmacological advances, (Burke, R.E. et al, 1981). Involuntary movements may occur in neurological disorders of many kinds, (Brett, E.M., 1983). In some disorders, however, involuntary movements of a dramatic sort constitute the main, if not the only, clinical abnormality.

Involuntary movements are seen in many disorders affecting the central nervous system, (Lockman, L.A., 1982). Clinically, these disorders consist not only of involuntary movements but also of abnormalities of posture and tone; changes in intellect and emotion may be present in varying degrees.

The term involuntary movement disorders implies that the movements are not under volitional control, the patient can not stop them at will. Also implicit in this term is the observation that the movements occur at rest, that is the absence of volitional movement. Furthermore, the movements are without apparent purpose.

The term extra pyramidal diseases also is used to refer to this group of conditions and suggests that they are the result of pathological changes outside the pyramidal tract, changes usually thought to occur in the basal ganglia: putamen, caudate, globus pallidus, substantia nigra, and subthalamic nucleus.

## The Extra pyramidal System

Bannister S.R. 1978, considered extra pyramidal syndromes as those disorders wich result from lesions involving those parts of the brain other than the corticospinal pathways which are concerned with movement. The principal such structures are those known as the basal ganglia, namely the corpus striatum and the nuclei which are anatomically and functionally associated with it.

The corpus striatum consists of the caudate nucleus and the lenticular nucleus, which is divided into the putamen and the globus pallidus. Walton, J.N. 1977, stated that the corpus striatum was phylogenetically, the oldest part of the cerebrum and it was lying in the substance of the cerebal hemisphere between the lateral ventricle and the insula. He also stated that it was consisting of the caudate nucleus and the lentiform nucleus, the latter being divided into the putamen and the globus pallidus.

Unfortunately, the structures included in the basal ganglia vary between workers, stated Williams, P.L. et al 1980, but most commonly the term includes the amygdaloid complex, the caudate nucleus and the lentiform nucleus. These topographical structures are a heterogenous group with respect to their structural and functional associations and phylogenetic history. Accordingly, a large and often confusing terminology has been generated by different investigators, the various structures being subdivided and regrouped in many different ways. The basal ganglia have been considered by Carpenter, M.B. 1976, as the principal component of the so called " extra pyramidal system." It has been used to conveniently group together an array of neural structures not otherwise easily designated. The tacit assumption has been that these structures functioned as a unit and formed a system. The term has been narrowed to include the corpus striatum, the subthalamic nucleus, the substantia nigra, the red nucleus, and the brain stem reticular formation - Ain Shams University

Carpenter M.B., 1978, divided the basal ganglia which is derived from the telencephalon into two major divisions a) the corpus striatum concerned with somatic motor functions, and b) the amygdaloid nuclear complex. Functionally related to the hypothalamus and regarded as an integral part of the Alimbic system.

a) The corpus striatum as described by Carpenter, M.B., 1978 is in close relationship to parts of the diencephalon, but separated from it by the internal capsule. It represents one of the Principal components of the basal ganglia. The corpus striatum consists of two distinctive parts, the globus pallidus and the neostriatum. The blobus pallidus is composed of medial and lateral segments oriented along the lateral surface of the internal capsule. This part of the corpus striatum is designated the paleostriatum, but commonly isreferred to simply as the pallidum. Arising as a single gray mass during early development, the corpus striatum becomes secondarily divided by the fibres of the internal capsule into two cellular masses, the lenticular nucleus and the caudate nucleus. This separation is incomplete. Rostroyentrally the head of the caudate nucleus is continuous with parts of the putamen. In more posterior regions portions of the caudate nucleus and dorsal parts of the putamen are connected a number of slender gray bridges between fibres of the internal capsule.

Carpenter, M.B. 1976, examined the Caudate nucleus and the putamen which were found to be cytologically identical. Cells are densely packed, exhibit no laminations or special groups and classically are considered to be of two types:

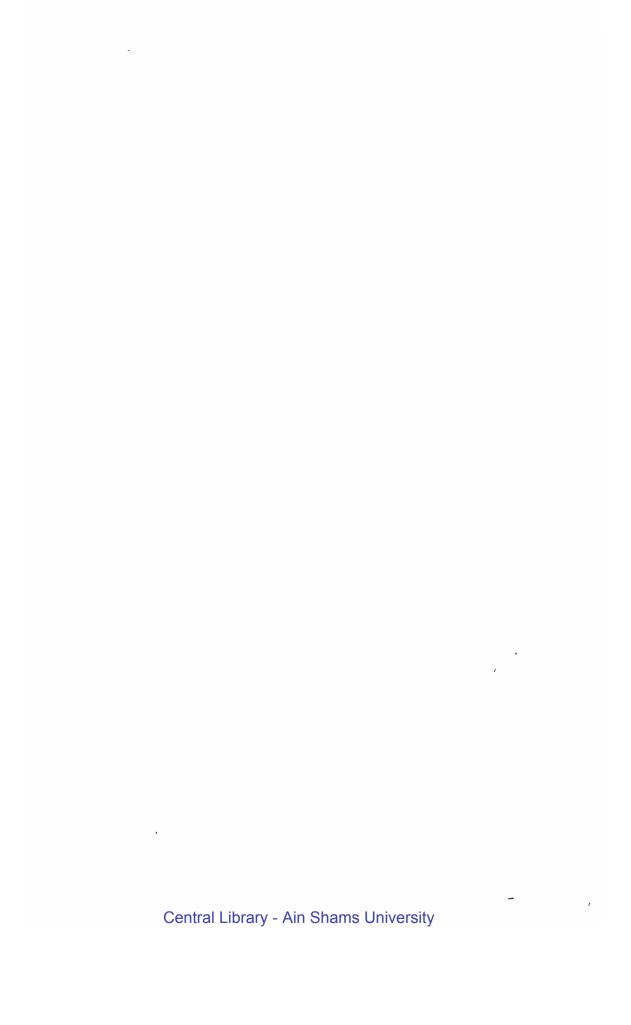
- 1- Small achromatic neurons and
- 2- Large multipolar neurons with rounded contour.

Williams, P.L. etal 1980, described the caudate nucleus as an arcuate mass of grey matter which has already been noted in Central Library - Ain Shams University

in the floor of the anterior cornu and central part of the lateral ventricle and the roof of its inferior cornu. Its anterosuperior end is massive and termed the head. At the interventrieular foramen this narrows into the body of the nucleus which tapers imperceptibly in to the tail. While Carpenter, M.B. 1978, described the caudate nucleus as an elongated arched gray mass related throughout its extent to the ventricular surface of the lateral ventricle. It's enlarged anterior end, or head, lies rostral to the thalamus and bulges into the anterior horn of the lateral ventricle. The body extends along the dorso lateral border of the thalamus from which it is separated by the stria terminalis and the terminal vein. This part of the caudate nucleus is regarded as suprathalamic. The tail of the caudate nucleus is the long, attenuated caudal portion which sweeps into the temporal lobe in the roof of the inferior horn of the lateral ventricle and comes into relationship with the central nucleus of the amygdaloid complex.

Williams, P.L. et al 1980, described the lentiform nucleus as a biconvex lens, but the curvature of its medial surface is sharper than the curvature of its lateral surface. It is completely buried in the substance of the hemisphere. Laterally it is covered by a thin layer of white matter which constitutes the external capsule. Medially the lentiform nucleus is in relation to the internal capsule which separates if from the thalamus behind and from the head of the caudate uncleus in front. Round its anterior superior and posterior margins the nucleus is related to the corna radiata.

Carpenter, M.B. 1978, added that a vertical plate white matter, the lateral medullary lamina, divides the nucleus into a larger outer portion, the putamen and an inner portion, the globus pallidus Central Library - Ain Shams University



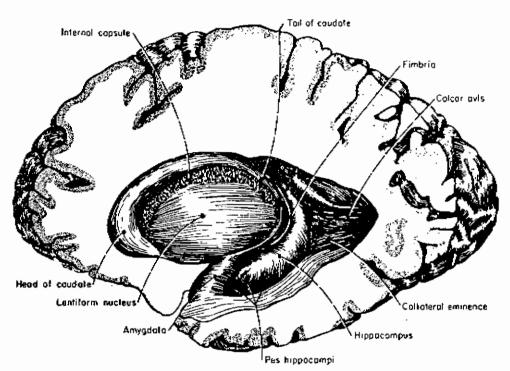


Fig. 108 A deep dissection of the lateral surface of the cerebral hemisphere to show the caudate and lentiform nuclei as well as the amygdala (amygdaloid nucleus)

- b)The putamen, the latgest and most lateral part of the basal ganglia, lies between the external copsule and the lateral medullary lamina of the globus pallidus. The caudate nucleus and the putamen, which are continous rostrally have essentially the same cytological structure.
- The globus pallidus as Walton,J.N.:1977, stated, jies medial to the putamen. It is separated from the thalamus and the caudate nucleus by the internal capsule which also separates the head of the caudate from the anterior part of the putmen.
- Carpenter, M.B. 1978, added that this nucleus forms the smaller and most medial segment of the lentiform nucleus. Throughout most of its extent the pallidum lies medial to the putamen, the internal capsule forms its medial border. A thin lateral medulary lamina is found on the external surface of the pallidum at its junction with the putamen, A medial medullary lamina divides the globus pallidus into medial and lateral segments.
- The claustrum is a thin plate of gray matter lying in the medullary substance of the hemisphere between the lenticular nucleus and the insular cortex which is separated from these structures by two white laminae the external capsule medially and the exterme capsule laterally. Although some consider the claustrum as a part of the striatum, it seems likely that it arises from the deeper layers of the insular cortex. Its function and connections remain obsure.

# Connections of the corpus striatum

In outline, (Williams, P.L. et al 1980), the neostriatum (putamen and caudate nucleus) constitutes the main receiving station, this projects to the globus pallidus, which in turn, Central Library - Ain Shams University

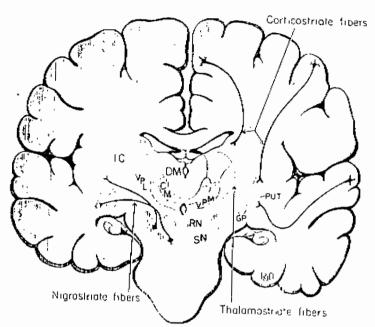


Fig. 11-8. Semischematic diagram of striatal afferent fibers. Corticustrate fibers (black) arising from broad cortical regions on the convexity of the hemisphere project to the patament three on the medial-surface projects largely to the caudate nucleus. Nigrostriatal fibers (red) arise from cells of the pars compacta. Thalamostriate fibers (blue) arise from the centromedian-parafascicular complex. All of these striatal afferent systems are topographically organized. (From Carpenter, Human Neuromatomy, 1976; courtesy of The Williams & Wilkins Company.)