LONG-TERM EEG MONITORING IN ACQUIRED EPILEPTIC DYSPHASIA

Thesis

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بسم الله الرحمن الرحيم

﴿قَالَ رب اشرح ِلي صدري ﴿ ويسر ِلي أمري
﴿ واحلُلْ عقْدةً من لسانِي ﴿ يفْقَهوا قَولي

صدق الله العظيم

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This effort is dedicated to

My Family

To whom I am originally, endlessly and proudly affiliated

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LIST OF ABBREVIATIONS

ADEM	:	Acute Disseminated EncephaloMyelitis
5-HT	:	Serotonin
ABR	:	Auditory Brainstem Response
AD	:	Autistic Disorder
ADHD	:	Attention Deficit Hyperactivity Disorder
AEA	:	Acquired Epileptic Aphasia
AEDS	:	Anti-Epileptic Drugs
AMT	:	Alpha-[^{11} C]Methyl-L-Tryptophan
ANOVA	:	ANalysis Of Variance
AR	:	Autistic Regression
AS	:	Asperger Syndrome
ASD	:	Autism Spectrum Disorders
BAEPs	:	Brainstem Auditory Evoked Potentials
BCEOP	:	Benign Childhood Epilepsy with Occipital Paroxysms
BCSSS	:	Benign Childhood Seizure Susceptibility Syndromes
BDNF	:	Brain Derived Neurotrophic Factor
BECTS	:	Benign Childhood Epilepsy with Centro-Temporal Spikes
BHTs	:	Behavioral Hearing Tests
BRE	:	Benign Rolandic Epilepsy
CAP	:	Cyclic Alternating Pattern
CAPD	:	Central Auditory Processing Disorder
CARS	:	Child Autism Rating Scale
CBC	:	Complete Blood Picture
CBCL	:	Child Behaviour CheckList
CBD	:	CorticoBasal Degeneration
CCTV-EEG	:	Closed-Circuit Television Videotaping and digitized EEG telemetry
CDKL5	:	Cyclin Dependent Kinase-Like 5
CFT	:	beta-Carbomethoxy-3 beta-4-Fluorophenyl Tropane
CHAT	:	CHecklist for Autism in Toddlers
CNS	:	Central Nervous System
CSF	:	Cerebro-Spinal Fluid
CSWS	:	Continuous Spikes-Waves during slow Sleep
CT	:	Computed Tomography
CTS	:	Centro-Temporal Spikes
dB	:	deci-Bell
DSM-IV	:	The Diagnostic and Statistical Manual of Mental Disorders, 4th Edition
DWI	:	Diffusion-Weighted Imaging
EA	:	Epileptiform Activity
EEG	:	Electroencephalogram
ELP4	:	Elongator Protein complex 4
EME	:	Early Myoclonic Encephalopathy
EMFs	:	Evoked Magnetic Fields
EPC	:	Epilepsia Partialis Continua
ERF	:	Event Related Field
EROs	:	Event Related Oscillations

ERPs	:	Event-Related brain Potentials
ESES	:	Electrical Status Epilepticus with Sleep
FDG	:	18-Fluoro-2-DeoxyGlucose
FISH	:	Fluorescence In Situ Hybridization
FMAER	:	Frequency-Modulated Auditory-Evoked Response
FMR1		Fragile-site Mental Retardation 1 gene
fMRI	:	functional MRI
FOS	:	Fixation Off Sensitivity
FXS	:	Fragile X Syndrome
GEFS+	:	Generalized Epilepsy with Febrile Seizures "plus"
GER	:	GastroEsophageal Reflux
GSES	:	Giant Somatosensory Evoked Spikes
HFA	:	High Functioning Autism
HIV	:	Human Immunodeficiency Virus
HMPAO	:	HexaMethylPropyleneAmine Oxide
HPA	:	Hypothalamic-Pituitary- Adrenal
ICOE-G	:	Idiopathic Childhood Occipital Epilepsy of Gastaut
IEP	:	Interictal eEpileptiform Discharges
IPL	:	InterPeak latencies
IQ	:	Intelligence Quotient
LFA	:	Low Functioning Autism
LGS	:	Lennox-Gastaut Syndrome
LKS	:	Landau Kleffner Syndrome
LTG	:	LaMoTrigine
MAE	:	Myoclonic-Astatic Epilepsy
MBDs	:	Methyl-CpG-Binding Domain proteins
MBP	:	Myelin Basic Protein
MCA	:	Middle Cerebral Artery
McDD	:	Multiple complex Developmental Disorder
MDI	:	MultiDimensionally Impaired disorder
MECP2	:	MEthyl-CpG binding Protein-2
MEG	:	MagnetoEncephaloGraphy
MMF	:	Magnetic Mismatch Field
MMN	:	MisMatch Negativity
MMR	:	Measles-Mumps-Rubella
MRI	:	Magnetic Resonance Imaging
MRS	:	Magnetic Resonanse Spectroscopy
MS	:	Multiple Sclerosis
MSNE	:	Myoclonic Status in Nonprogressive Encephalopaties
NCG	:	Normal Control Group
NGF	:	Nerve Growth Factor
NREM	:	Non–Rapid Eye Movement
OCD	:	Obsessive-Compulsive Disorder
PDA	:	Pathological Demand Avoidance
PDD-NOS	:	Pervasive Developmental Disorder – Not Otherwise Specified
PDDs	:	Pervasive Developmental Disorders
PET	:	Positron Emission Tomography

PLEDs	:	Periodic Lateralized Epileptiform Discharges
PS	:	Panayiotopoulos Syndrome
PSG	:	PolySomnoGraphy
PWI	:	Perfusion Weighted Imaging
QEEG	:	Quantified EEG
rCBF	:	regional Cerebral Blood Flow
RD	:	Reading Disability
RE	:	Rolandic Epilepsy
REM	:	Rapid Eye Movement
RS	:	Rett Syndrome
SB	:	Suppression Burst
SD	:	Standard Deviation
SD	:	Sudden Death
SLI	:	Specific Language Impairment
SNRIs	:	Nonselective Serotonin Reuptake Inhibitors
SPECT	:	Single-Photon Emission Computed Tomography
SPGI	:	Superior tip of the Precentral Gyrus of the Insula
SPL	:	Sound Pressure Level
SPS	:	Semantic-Pragmatic Syndrome
SQ	:	Social Quotient
SSD	:	Speech Sound Disorder
SSR	:	Steady-State Responses
SSRIs	:	Selective Serotonin Reuptake Inhibitors
STG	:	Superior Temporal Gyrus
STK9		Serine Threonine Kinase 9
STS	:	Superior Temporal Sulcus
SW	:	Slow Wave
SW	:	Spike-Wave
SWS	:	Slow Wave Sleep
TBI	:	Traumatic Brain Injury
TORCH	:	TOxoplasmosis, Rubella, Cytomegalo virus, Herpes simplex virus
USA	:	The United States of America
VABS	:	Vineland Adaptive Behaviour Scales
VNS	:	Vagal Nerve Stimulator

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ABSTRACT

INTRODUCTION: Dysphasia is a language disorder in which there is an impairment of speech and of comprehension of speech. Acquired childhood aphasia is rare but has important conceptual implications for developmental neuropsychology. **AIM:** Studying long term monitoring EEG in patients with acquired dysphasia, whether giving history of epilepsy or not, to stand on the different EEG pictures related to it, identify their causes and calculate their ratios. **SUBJECTS and METHODS:** 40 patients, males and females, with age ranging 3-16 years, giving history of acquired language disorder were studied for 1. Long-term, sleep and wake EEG, 2. Psychometric assessment; Childhood behavior checklist (CBCL), Childhood autistic assessment scale (CARS) and Vineland Adaptive Behaviour Scales (VABS), 3. Auditory evoked potential studies: early (BAEPs) and –whenever possible-late (P300). In addition, 20 normal age and sex matched subjects were examined for; 1. Auditory evoked potential studies, 2. CBCL and VABS. **RESULTS:** The patient group (24 males and 16 females), were divided to group I giving history of epilepsy or having epileptiform EEG and group II negative for either. Group I showed heterogeneous EEG pictures, and heterogeneous diagnoses, yet 7 patients showed sleep induced potentiation for epileptiform changes. Group II patients chiefly had autism spectrum disorder. Statistical comparisons and correlations were performed for patient groups and controls. **Conclusion:** The boundaries between epileptiform EEG, epilepsy and epileptic encephalopathy are clear by definition, but they usually merge in clinical cases. The onset of epilepsy in brain systems involved in social communication and/or recognition of emotions- during the brain developmental period- can initiate speech and language development impairment. Also, it can occasionally be the cause of -or may aggravate preexisting behavioural disturbances.

Keywords:

Long-term
EEG monitoring
Acquired epileptic dysphasia
Neurophysiology
Neuropsychology

INTRODUCTION

AND

AIM OF WORK

"Speak! That I May See You"
Socrates

Definition of dysphasia:

Dysphasia is a language disorder in which there is impairment of speech and of comprehension of speech. It is caused by brain damage, usually in the left side of the brain which is responsible for language and communication. The word comes from the Greek *dys-* (impairment) and *phasia* (speech). The term dysphasia has been eclipsed by the modern usage of the term "aphasia" particularly in the field of speech/language pathology so as not to confuse with the swallowing disorder "dysphagia". Aphasia literally means no speech. But the speech impairment in aphasia could range from complete absence of speech to difficulty in naming a few objects. *(http://en.wikipedia.org/wiki/Dysphasia)*

Acquired childhood aphasia is rare but has important conceptual implications for developmental neuropsychology *(Van Hout, 1997).*

Defferential diagnosis of Acquired Epileptic Aphasia (AEA):

- **Acute Disseminated Encephalomyelitis**

Acute disseminated encephalomyelitis (ADEM) is a monophasic, demyelinating disease of the central nervous system that predominately affects prepubertal children. It is typically characterized by an abrupt onset of neurologic symptoms preceded by an infection or recent immunization *(Harris and Lee, 2007).*

- **Benign Childhood Epilepsy**

Benign childhood epilepsy with centro-temporal spikes (BECTS) is the most common form of focal idiopathic epilepsy in childhood, showing no demonstrable anatomical lesions. In general the prognosis is good and the seizures disappear by 15 years of age, with normalization of the electroencephalogram (EEG) *(Fonseca et al, 2007).*

- **Cardioembolic Stroke**

-**Epilepsy in Children with Mental Retardation**

-**Epileptic and Epileptiform Encephalopathies**

Epileptic encephalopathies are progressive clinical and electroencephalographic syndromes where deterioration is thought to be caused by frequent seizures and abundant EEG epileptiform activity *(Tharp, 2004).*

- **Head Injury**

- **Low-Grade Astrocytoma**

Low-grade astrocytomas are a heterogeneous group of intrinsic central nervous system (CNS) neoplasms that share certain similarities in their clinical presentation, radiologic appearance, prognosis, and treatment *(Jallo and Benardete, 2007).*

- **Mental Retardation**

- **Neurocysticercosis**

The most common helminthic disease of the nervous system and currently represents a major public health problem in developing countries of Latin America, Asia, and Africa, as well as in industrialized nations with a high immigration rate of people from endemic areas *(Del Brutto, 2005)*

Other Problems to be Considered:

-Acquired expressive epileptic aphasia

-Adrenoleukodystrophy

-Childhood disintegrative disorder

-Developmental dysphasia or developmental expressive language disorder

-Disintegrative epileptiform disorder

- Electrical status epilepticus with sleep (ESES) *(De Menezes, 2010).*

Landau Kleffner Syndrome

LKS typically develops in healthy children who acutely or progressively lose receptive and expressive language ability coincident with the appearance of paroxysmal EEG changes. In 1957, Landau and Kleffner initially described acquired epileptic aphasia (AEA) and subsequently reluctantly agreed to the attachment of their names to the syndrome *(De Menezes, 2010).*

Landau-Kleffner syndrome is characterized by acquired aphasia and paroxysmal, sleep-activated EEG paroxysms predominating over the temporal or parieto-occipital regions. Secondary symptoms include psychomotor or behavioral disturbances and epilepsy with a favorable outcome for seizure control. The prevalence is unclear. A male predominance exists, with an approximately 2:1 ratio. This regressive syndrome affects children after having achieved early developmental milestones, with 3–9 years being the usual age of presentation *(Pearl et al., 2001)*