Guillain Barré syndrome: subtypes, treatment effect and prognosis: A retrospective study in an Egyptian pediatric intensive care unit

Thesis

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By

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Abstract

Background: Guillain-Barré syndrome (GBS) is mostly an acute inflammatory demyelinating ascending polyradiculoneuropathy.

Aims: to estimate 1) number, age and sex variations of GBS patients, who were admitted to the Cairo University Pediatric Intensive Care Unit in a five-year retrospective study, 2) incidence of other acute flaccid paralysis mimicking GBS, 3) antecedent illnesses preceding GBS and 4) electrophysiological patterns, clinical variants and response to treatment of GBS patients.

Methods: this is a retrospective study of all children with acute flaccid paralysis admitted to Cairo University Pediatric Intensive Care Unit between June 2009 and June 2014**.**

Results: This study detected 52/61 cases (85.2%) had GBS. This study detected 10/52 cases (19.2%) were in the first year age group, 30/52 cases (57.7%) were in the age group from 2-5 years and 12/52 cases (23.1%) were in the age group from 6-12 years. This study detected 27/35 upper respiratory infection cases (77.1%) had GBS, 21/21 gastroenteritis cases (100%) had GBS. This study detected 27/52 cases (44.3%) having acute inflammatory demyelinating polyneuropathy and 17/52 cases (29.5%) having acute motor axonal neuropathy. Miller Fisher syndrome was associated in 5/52 cases (8.2%) and Bickerstaff encephalitis was associated in 4/52 cases (6.6%). Improvement occurred in 47/52 cases (90.4%) and 5/52 cases (9.6%) showed slow improvement and prolonged stay.

Conclusions: GBS was the commonest cause of acute flaccid paralysis. The most commonly affected age group was from 2-5 years. It appears that acute inflammatory demyelinating polyneuropathy was the most common subtype in our study. High percentage of GBS cases had favorable outcome among our study group.

Keywords: Guillain-barré syndrome – Acute inflammatory demyelinating polyneuropathy – Acute flaccid paralysis.

Abbreviations

AChR	Acetylcholine receptor
AIDS	Acquired immune deficiency syndrome
AFP	Acute flaccid paralysis
AIDP	Acute inflammatory demyelinating polyneuropathy
AMAN	Acute motor axonal neuropathy
AAN	American Academy of Neurology
anti-AChR Abs	Anti acetylcholine receptor antibodies
AD	Autonomic disturbances
BBE	Brain stem encephalitis
CUSPH	Cairo University Pediatric Intensive Care Unit
C.Jejuni	Campylobacter Jejuni
CDC	Centers for Disease Control and Prevention
CNS	Central nervous system
CSF	Cerebrospinal fluid
CIDP	Chronic inflammatory demyelinating polyneuropathy
C. botulinum	Clostridium botulinum
C	Complement
CMAP	Compound Muscle Action Potentials
CT	Computed tomography
CS	Corticosteroids
C diphtheria	Corynebacterium diphtheria
CK	Creatine Kinase

CIM	Critical illness myopathy
CIP	Critical illness polyneuropathy
CMV	Cytomegalovirus
cDNA	DNA complement
EDx	Electrodiagnostic studies
EEG	Electroencephalography
EMG	Electromyography
ELISA	Enzyme-linked immunosorbent assay
GBS	Guillain-Barre Syndrome
HFMD	Hand, foot, and mouth disease
HIV	Human immunodeficiency virus
HLA	Human leucocyte antigen
HypoPP	Hypokalemic periodic paralysis
IVIg	Intravenous Immunoglobulin
JEV	Japanese encephalitis virus
MRI	Magnetic resonance imaging
MAC	Membrane-attack complex
MFS	Miller Fisher Syndrome
MuSK	Muscle-specific tyrosine kinase
MG	Myasthenia gravis
NCS	Nerve conduction studies
NMJ	Neuromuscular junction
NSAIDS	Nonsteroidal anti-inflammatory drugs
PNS	Peripheral nervous system
PE	Plasma Exchange
PSGBS	Plasma Exchange Sandoglobulin Guillain-Barre’
PCR	Polymerase chain reaction

PM	Polymyositis
K+	Potassium
RFFIT	Rapid fluorescent focus inhibition test
RT-PCR	Reverse transcriptase-polymerase chain reaction
Na+	Sodium
SIADH	Syndrome of inappropriate antidiuretic hormone
US	United States
VZV	Varicella-zoster virus

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Introduction

Acute flaccid paralysis (AFP) is defined as sudden onset of weakness and floppiness in any body part in a child aged less than 15 years or paralysis in a person of any age in which polio is suspected **(Alexander et al., 1997).** The most frequent cause of AFP that must be distinguished from Poliomyelitis is GBS. The paralysis of GBS is typically symmetrical, and maybe associated with sensory changes. Fever, nausea, headache, vomiting and pleocytosis are usually absent in GBS. Other important causes of AFP include transverse myelitis, traumatic neuritis, infectious and toxic neuropathies. Non polio enteroviruses like Coxsackie A, Coxsackie B, ECHO or Enterovirus types 70 and 71 have also been temporally associated with AFP and most of these cases show a course of improvement with complete recovery **(Agrawal and Singh, 2004).**

Guillain-Barré syndrome (GBS) is mostly an acute inflammatory demyelinating ascending polyradiculoneuropathy **(Jasem et al., 2013).** Guillain-Barré syndrome is, currently, the most common cause of acute flaccid paralysis following the worldwide decline in the incidence of poliomyelitis. Incidence varies according to age, geographic areas and diagnostic criteria used for inclusion. Annual incidence in western countries varies from 1.1 to 1.8/100,000 population per year with a considerably lower annual incidence of 0.66/100,000 population per year in each of Taiwan and China **(El-Bayoumi et al., 2011).**

Flu-like illness or gastroenteritis precedes the onset of paralysis by 6 weeks in about two-thirds of patients. The culprit infectious agent often remains unrecognized, but Campylobacter

jejuni Mycoplasma pneumonia, and Cytomegalovirus are commonly reported triggering pathogens. Molecular mimicry between structural components of both pathogens and myelin sheath of peripheral nerves, with subsequent cross-reaction of antibodies with the latter, is a commonly proposed hypothesis for the pathogenesis of disease **(Jasem et al., 2013).** GBS can be subdivided into the acute inflammatory demyelinating polyneuropathy (AIDP), the most frequent form in the western world; acute motor axonal neuropathy (AMAN), most frequent in Asia and Japan; and in Miller-Fisher syndrome (MFS), much more common in Japan than in the United States **(Anthony et al., 2012).** Additionally, overlap syndromes exist (GBS-MFS overlap) **(Van Doorn , 2013).**

Distal paresthesias evolve into symmetric progressive ascending areflexic motor weakness often in association with facial weakness and pain in limbs and back. Weakness may progress rapidly, necessitating the need for ventilatory support and may be associated with autonomic dysfunction **(Kannan et al., 2011).** Both intravenous immunoglobulin (IVIG) and plasma exchange (PE) are effective in GBS. Rather surprisingly, steroids alone are ineffective **(Van Doorn, 2013).** The routine use of IVIG as the first line of treatment in GBS followed the publication of a randomized controlled trial in 1992 showing a similar, if not a superior, effect of IVIG compared to PE **(El-Bayoumi et al., 2011).**

Aim of work

- To estimate number, age and sex variations of GBS patients, who were admitted to the Cairo University Pediatric Intensive Care Unit in a five-year retrospective study.
- To estimate incidence of other acute flaccid paralysis mimicking GBS.
- To estimate antecedent illnesses preceding GBS.
- To estimate electrophysiological patterns, clinical variants and response to treatment of GBS patients.

Guillain-Barre Syndrome (GBS)

Acute flaccid paralysis:

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Guillain-Barré Syndrome (post infectious polyneuropathy)

a. Definition

The French physician Jean Landry (1826-1865), first described the disorder in 1859. In 1916, Georges Guillain (1876-1961), who was a French neurologist and Jean Alexandre Barré (1880-1967), who was a French neurologist, and André Strohl André Strohl (1887- 1977), who was a French physiologist diagnosed two soldiers with the illness **(Wals et al., 2012).** Since the eradication of polio in most parts of the world, GBS has become the most common cause of acute flaccid paralysis **(El-Bayoumi et al., 2011).** GBS is an autoimmune disorder of the peripheral nervous system (PNS), characterized by weakness, usually symmetrical, evolving over a period of several days or more. Affected persons rapidly develop weakness of the limb, weakness of the respiratory muscles and areflexia. The post infection polyneuropathy that causes