



Guillain-Barre Syndrome, Familial

Alternative Names

GBS
Polyneuropathy, Inflammatory Demyelinating,
Acute
AIDP
Polyneuropathy, Inflammatory Demyelinating,
Chronic
CIDP

Record Category

Disease phenotype

WHO-ICD

Diseases of the nervous system > Polyneuropathies
and other disorders of the peripheral nervous
system

Incidence per 100,000 Live Births

2-5

OMIM Number

139393

Mode of Inheritance

N/A

Gene Map Locus

N/A

Description

Guillain-Barre syndrome is a rare immune-mediated acute polyneuropathy that occurs in previously healthy individuals and may lead to a variety of motor and sensory deficits. The clinical spectrum of Guillain-Barre syndrome is heterogenous and encompasses acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy, acute motor and sensory axonal neuropathy, and Miller Fisher syndrome. The disease is characterized by a rapid onset of symmetrical limb weakness, which progresses over days to four weeks. In the majority of cases, Guillain-Barre syndrome has been associated with antecedent *Campylobacter jejuni* infections. The pathogenesis of *C. jejuni*-induced Guillain-Barre syndrome is complex and probably involves unique

virulence factors associated with the organism, as well as the host genetic susceptibility factors.

Campylobacter strains with certain Penner heat-stabile serotypes, including HS:1, HS:2, HS:4, HS:4/50, HS:5, HS:10, HS:13/65, HS:16, HS:19, HS:23, HS:35, HS:37, HS:41, HS:44, and HS:64, have been reported to be overrepresented among isolates from Guillain-Barre syndrome cases. Several studies indicate that HS:19 and HS:41 have a clonal population structure.

Molecular Genetics

There is evidence that some cases of inflammatory demyelinating polyneuropathy may be caused by mutation in the PMP22 gene on chromosome 17.

Epidemiology in the Arab World

Jordan

Abu-Haweleh and Hiyari (1998) undertook a retrospective study of 18 Jordanian children (ten males, eight females; mean age: 5.8 years), admitted between 1988 and 1996 with GBS, to estimate the neurological sequela of the disease. Two peaks of incidences were noted; one below the age of 3-years and one from 9-11 years. In terms of clinical features, all patients were found to have a flaccid ascending symmetrical paralysis as well as absent to +1 deep tendon reflexes. Other features seen in patients were sensory complaints (55%), cranial nerves dysfunction (44%; facial nerve being the most commonly involved), autonomic disturbances (4%), and respiratory failure (2%). One patient each had diabetes insipidus and abnormal behavior. Nerve conduction studies showed demyelination of varying intensity in all patients. Nine of the patients (50%) had a history of previous infection; 33% having upper respiratory tract infections. In most cases, the period to peak paralysis was about two weeks. Only one of the patients, a six-year old boy, died. He had a history of gastroenteritis 10 days prior to the weakness. He was stable on mechanical ventilation, but arrested suddenly, possibly due to cardiac arrhythmia. Three other patients were left with residual weakness two years after the onset of the disease. The morbidity



and mortality rates were, therefore, 16% and 5.6%, respectively. Abu-Haweleh and Hiyari (1998) found no definite correlation between clinical features and prognosis for GBS among these patients, but commented that both patients who required ventilatory support had a poor prognosis; one dying and the other left with a motor disability.

Kuwait

Nagarajan and Al-Shubaili (2006) conducted a study to examine the clinical and neurophysiological pattern of Guillain-Barre syndrome on 41 patients (including 21 Kuwaitis) attending Ibn Sina Hospital. The mean age was 41 ± 15.9 years and the male to female ratio was 2.7:1, while the Kuwaiti male to female ratio was 1.8:1. The dominant clinical presentation was the proximal lower limb (LL) weakness. Nerve conduction studies (NCS) revealed a demyelinating pattern in 70%, an axonal pattern in 15%, mixed type in 5% and no abnormality in the remaining 5%. Nagarajan and Al-Shubaili (2006) demonstrated that the majority of subjects (70%) improved using one course of intravenous immunoglobulin (IV IG) with a mean recovery time (MRT) of 4.4 weeks. While GBS patients with predominant distal weakness in the LL, proximal weakness in the upper limb (UL), autonomic disturbance, and axonal type GBS showed a delayed recovery with MRT: 7.8, 6, 6.5, and 6 weeks, respectively in the NCS. Nagarajan and Al-Shubaili (2006) suggested that patients possessing any of these characteristics should undergo immunotherapy early in the course of the disease.

Vurgese et al. (2006) described a 35-year old Arab patient who presented with fleeting pain and tingling sensations in all limbs, and mild weakness in the lower limbs. He had a history of admission with complaints of syncope and palpitations. Three of his siblings and a first-degree relative had died suddenly after complaining of syncope and palpitation. Examination revealed mild hypotonia in the lower limbs, positive mycoplasma serology, elongated Q-T interval at 505 msec, elevated cardiac marker Troponin I, and abnormal CSF glucose level (3.76 mmol/L). Nerve conduction studies showed slowing of conduction, prolongation of distal latencies and F-wave, and conduction blocks suggestive of demyelinating polyneuropathy. The observations prompted Vurgese et al. (2006) to make the extremely rare diagnosis of Guillain Barre syndrome in comorbidity with Romano Ward syndrome. A recent *Mycoplasma* infection had probably triggered the GBS. The patient was closely monitored and treated with IV immunoglobulin for five days, and discharged without any neurological

deficit. He was advised to get an automated cardioverter defibrillator implanted.

Oman

Koul et al. (2003) conducted a prospective study to determine the outcome of patients with Guillain-Barre syndrome who received intravenous immunoglobulin and compared it with those who did not receive such a management. The data of all patients confirmed to have this disease were recorded. Severity of the disease was assessed clinically. All patients received intravenous immunoglobulin, 400mg/kg/dose for five doses (one patient received plasmapheresis as well), and in case of relapse, three additional intravenous immunoglobulin doses were given. In the study period of 10 years (1992 to 2001), 42 cases (24 males and 18 females) were admitted with Guillain-Barre syndrome with a mean age at presentation of 4.6 years, and mean onset of weakness of 8.9 days prior to admission. In 26 patients (61.9%), there were preceding events, with upper respiratory tract infection in 21 and immunization-related illness in three. Clinical examination revealed meningeal signs in 13 patients (30.5%), cranial nerve involvement in 23 patients (facial nerve in 19, and third and sixth nerve palsies in three), bulbar palsies in 6 patients (14.3%), and autonomic nervous system involvement in three (all had hypertension but tachycardia in one). Cerebrospinal fluid analysis in 39 patients revealed albuminocytology dissociation in 38. Upon comparison of the CSF cytology and biochemistry between the two groups of patients with and without meningeal signs, no difference was detected. Nerve conduction studies revealed normal sensory conduction in four and abnormal motor nerve conduction in 28 patients (66.7%). None of the patients had an acute motor axonal neuropathy. The mean recovery after receiving the intravenous immunoglobulin was within 5.8 days, but within six to 21 days from the first sign of recovery, five children (11.9%) relapsed and four of them responded to the repeat intravenous immunoglobulin doses while one required ventilatory support. During the course of the disease, the mean hospital stay was 20.4 days with seven children requiring ventilation for a mean of 27.9 days, and complete recovery was noted after a mean of 73 days (45 to 282 days). Residual deficit was noted in two children as they continued to have bilateral foot drop even after two years of follow up, but in this study group there were no mortalities. When this study was compared with the retrospective data, children who had received immunoglobulin had less hospital duration stay, ventilatory requirement and mortality rates, but similar rates of residual deficit when compared to those who did not receive immunoglobulin, but in



the prospective study, there was a relapse rate of 11.9% which was absent in the retrospective study.

United Arab Emirates

Engberg et al. (2001) analyzed strains of *Campylobacter jejuni* from different worldwide geographic locations involved in Guillain-Barre syndrome to determine serotype-independent epidemiologic marker in Guillain-Barre syndrome strains other than HS:19. Among the 11 patients with non-HS:19 Guillain-Barre syndrome-related C. strains one was from the United Arab Emirates. The HS:4' strain of the Emirati patient was identical to that in a Danish subject with Guillain-Barre syndrome and both shared a profile with a different HS:4' strain described in another Danish subject with diarrhea. Engberg and colleagues realized that the results in these subjects as well as in all other studied patients show that the isolates represent a heterogenic population and do not constitute a unique population across serotypes.

References

Abu-Haweleh A, Hiyari M. Neurological sequelae of guilain-barre syndrome in Jordanian children. Qatar Med J. 1998; 7(1):31-3.
Engberg J, Nachamkin I, Fussing V, McKhann GM, Griffin JW, Piffaretti JC, Nielsen EM, Gerner-Smidt P. Absence of clonality of *Campylobacter jejuni* in serotypes other than HS:19 associated with Guillain-Barre syndrome and gastroenteritis. J Infect Dis. 2001; 184(2):215-20. PMID: 11400076

Koul R, Chacko A, Ahmed R, Varghese T, Javed H, Al-Lamki Z. Ten-year prospective study (clinical spectrum) of childhood Guillain-Barre syndrome in the Arabian peninsula: comparison of outcome in patients in the pre- and post-intravenous immunoglobulin eras. J Child Neurol. 2003; 18(11):767-71. PMID: 14696904

Nagarajan V, Al-Shubaili A. Clinical and neurophysiological pattern of Guillain-Barre syndrome in Kuwait. Med Princ Pract. 2006; 15(2):120-5. PMID: 16484839

Vurgese T, Mapkar OA, Surrin SK. Guillain-Barre syndrome in a patient with Romano-Ward syndrome: a case report. Kuwait Med J. 2006; 38(1):53-5. [FT]

Related CTGA Records

Long QT Syndrome

External Links

<http://www.gbs.org.uk/index2.shtml>
<http://www.guillain-barre.com/overview.html>
http://www.ninds.nih.gov/health_and_medical/disorders/gbs.htm
<http://www.orpha.net/data/patho/GB/uk-Guillain.pdf>

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