XI CURSO DE ENFERMEDADES MUSCULARES
EN LA INFANCIA Y ADOLESCENCIA
MADRID 27 marzo, 2014

SINDROME DE GUILLAIN BARRE ASPECTOS CLINICOS Y TERAPEUTICOS

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Síndrome de Guillain-Barré

➢DEFINICION

- enfermedad autoinmune,
- desencadenada por una infección viral o bacteriana,
- caracterizada por una debilidad simétrica, rápidamente progresiva, de comienzo distal y avance proximal, a veces llegando a afectar la musculatura bulbar respiratoria,
- con pérdida de reflejos osteotendinosos
- con signos sensitivos leves o ausentes
- El LCR muestra una disociación albúmino-citológica, con aumento de proteinas y normalidad celular.

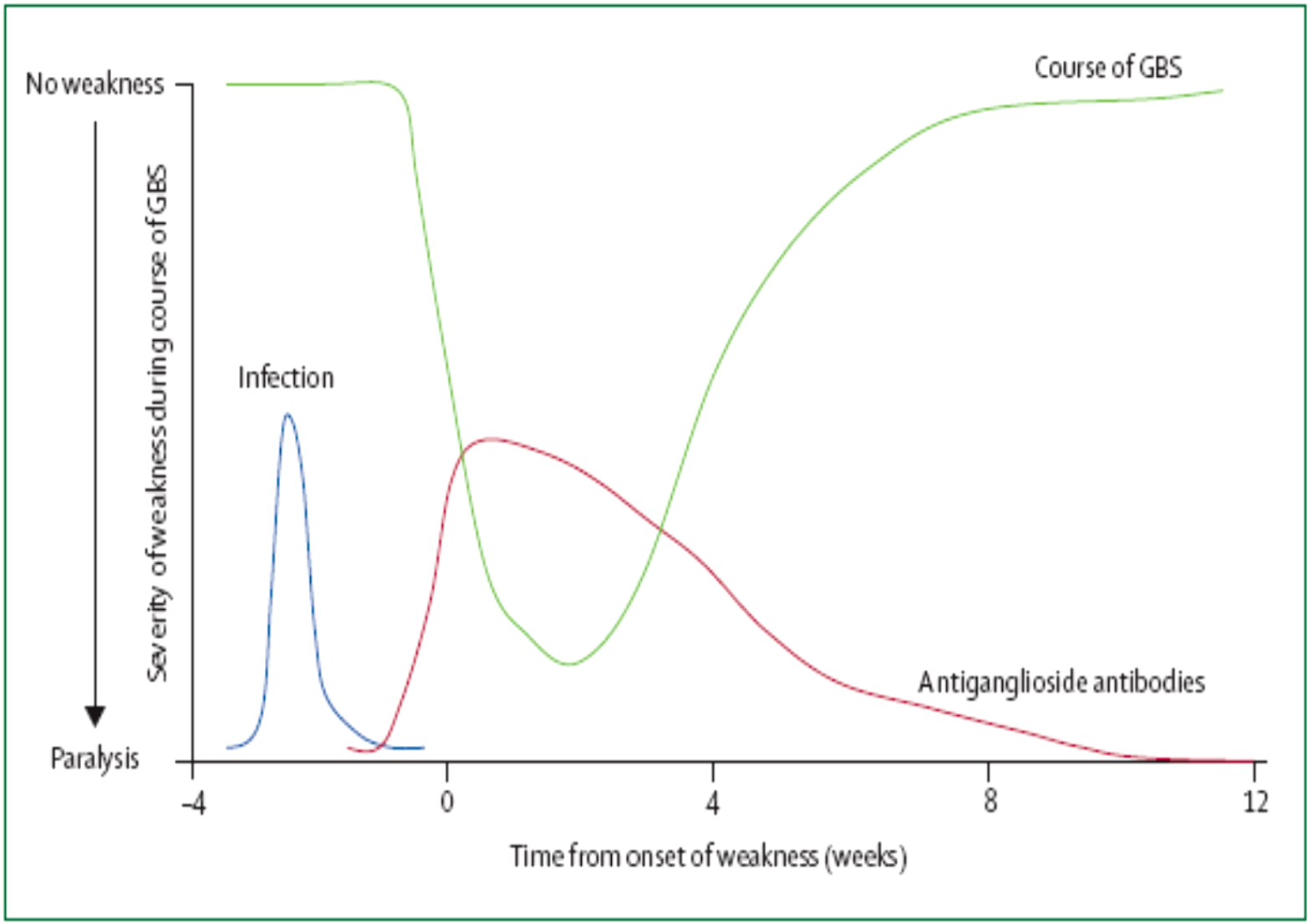


Figure 1: Relation between infections, antiganglioside antibodies, and clinical course of GBS

Van Doorn and Ruts.- Lancet Neurol 2008; 7:939-950

Síndrome de Guillain-Barré

Incidencia anual 1,2-2,3/100 mil, con aumentos temporales

Infecciones precedentes: 2/3 de los casos.

Los gérmenes causantes más frecuentes:

1)**Campylobacter jejuni** (26-41% de los casos). Está asociado especialmente a formas axonales y al síndrome de Miller-Fisher. Se puede aislar en las heces hasta varias semanas tras la terminación de la diarrea.
2) **Citomegalovirus** (10-22%).
3) **Epstein-Barr** (10%).
4) **Hemophylus influenzae** (2-13%)
5) **Varicela-zoster**
6) **Mycoplasma pneumoniae.**

Síndrome de Guillain-Barré.

CRITERIOS DE DIAGNOSTICO

Adaptados de Asbury y Cornblath, 1990.

I.- Criterios requeridos para el diagnóstico

II.- Rasgos que apoyan fuertemente el diagnóstico

- **Clinicos**
- **LCR**
- **Electrofisiológicos**

III.- Rasgos que hacen el diagnóstico dudoso.

IV.- Rasgos que descartan el diagnóstico

Síndr. de Guillain-Barré.

CRITERIOS DE DIAGNOSTICO

Adaptados de Asbury y Cornblath, 1990.

I.- Criterios requeridos para el diagnóstico

A.- **Debilidad progresiva** en más de un miembro.

B.- **Arreflexia osteotendinosa** universal.

Síndr. de Guillain-Barré.

CRITERIOS DE DIAGNOSTICO, cont.

II.- Rasgos que apoyan fuertemente el diagnóstico

A.- Rasgos clínicos (por orden de importancia):

1.- Progresión de la debilidad.
2.- Afectación relativamente simétrica.
3.- Síntomas y signos sensitivos leves.
4.- Afectación de nervios craneales.
5.- Recuperación. Comienza tras 2-4 semanas.
6.- Disfunción autonómica (taquicardia, hipotensión postural, hipertensión arterial, signos vasomotores).
7.- Ausencia de fiebre al comienzo.

Rasgos clínicos variantes (no van en orden de importancia):

1.- Fiebre al comienzo.
2.- Pérdida sensorial severa, con dolor.
3.- Progresión más allá de 4 semanas.
4.-Cese de la progresión sin recuperación o con secuelas permanentes importantes.
5.- Afectación de esfínteres.
6.- Afectación del SNC.

Síndr. de Guillain-Barré.

CRITERIOS DE DIAGNOSTICO, cont.

II.- Rasgos que apoyan fuertemente el diagnóstico, cont.

B.- Criterios de LCR:

1.- Proteinas aumentadas tras la 1ª semana.
2.- 10 células/mm3 o menos (leucocitos mononucleares).

Variantes:

1.- Sin aumento de proteinas en LCR en 1-10 semanas (raro).
2.- LCR con 11-50 leucocitos mononucleares.

Síndr. de Guillain-Barré.

CRITERIOS DE DIAGNOSTICO, cont.

II.- Rasgos que apoyan fuertemente el diagnóstico, cont.

C.- Criterios electrofisiológicos

80% tienen disminución de la velocidad de conducción.
Las latencias distales están aumentadas.
Abolición o retardo de la latencia de onda F.

Pero un 20% pueden tener normal la VC, y con frecuencia puede tardar en enlentecerse.

Síndr. de Guillain-Barré.

CRITERIOS DE DIAGNOSTICO, cont.

III.- Rasgos que hacen el diagnóstico dudoso.

1.- Asimetría marcada o persistente de la afectación.
2.- Disfunción vesical o rectal marcada.
3.- Disfunción vesical o rectal presentes al comienzo.
4.- Más de 50 leucocitos mononucleares en LCR.
5.- Presencia de leucocitos polinucleares en el LCR.
6.- Nivel sensitivo nítido, agudo.

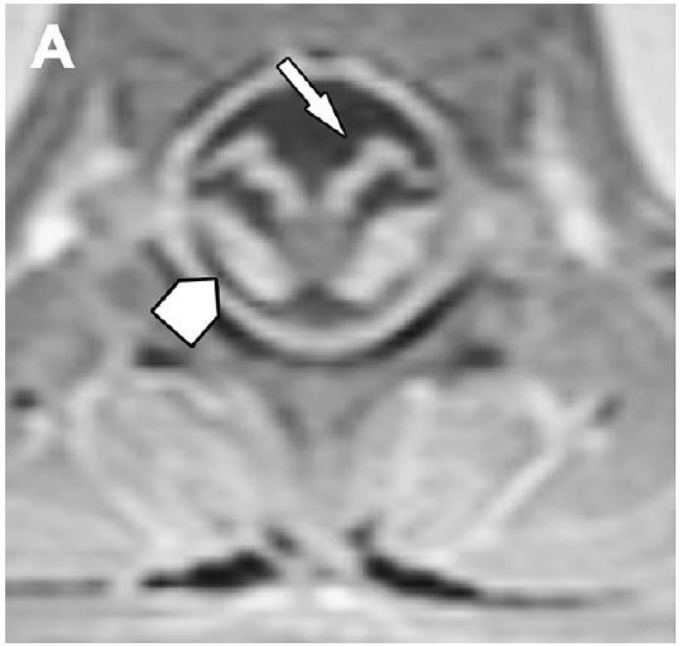
IV.- Rasgos que descartan el diagnóstico

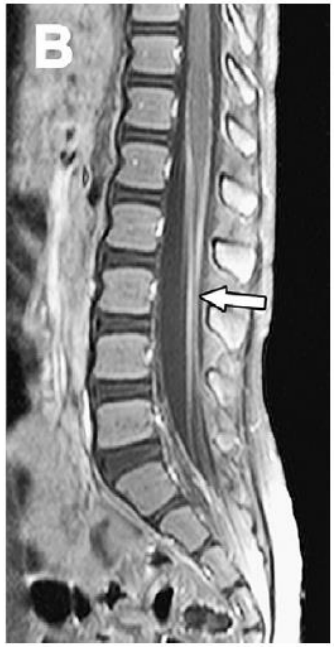
1.- Intoxicación por hexacarbonados, Porfiria aguda intermitente, difteria, neuropatía por plomo, poliomielitis, botulismo, parálisis histérica, neuropatía tóxica.
2.- Síndrome sensitivo aislado.
3.- Progresión de la afectación durante más de 2 meses (se trataría de una Poliradiculoneuropatía crónica inflamatoria desmielinizante).

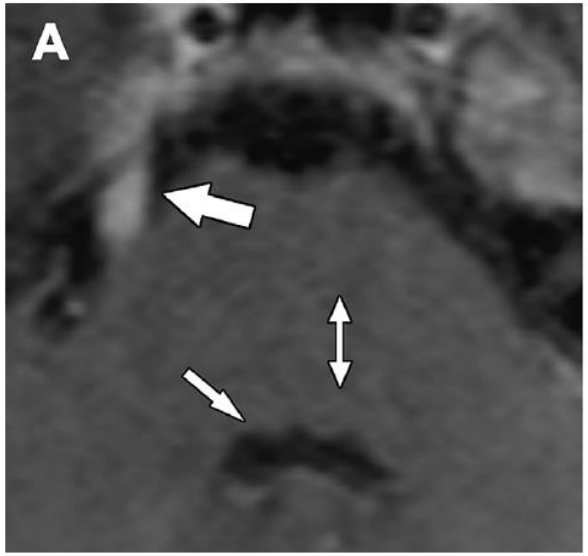
Nerve Root Enhancement on Spinal MRI in Pediatric Guillain-Barré Syndrome

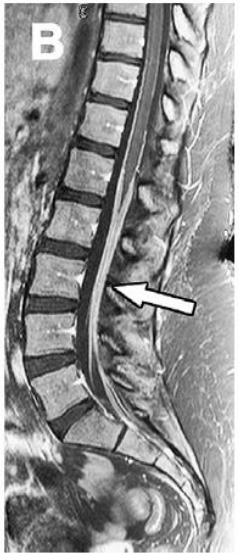
Sarah B. Mulkey, MD*, Charles M. Glasier, MD†, Bassem El-Nabbout, MD‡,
William D. Walters, MD, MPH*, Christian Ionita, MD*, Michael H. McCarthy, BA*,
Gregory B. Sharp, MD*, and Rolla M. Shbarou, MD*

Pediatr Neurol 2010;43:263-269.









Nerve Root Enhancement on Spinal MRI in Pediatric Guillain-Barré Syndrome

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We evaluated the frequency of nerve root enhancement on spinal magnetic resonance imaging in children with Guillain-Barré syndrome. At a single tertiary pediatric center, we conducted a retrospective chart review of children with Guillain-Barré syndrome who had complete spinal or lumbosacral spinal magnetic resonance imaging with gadolinium administration from January 2002-January 2009. Twenty-four consecutive patients were identified. Spinal nerve root enhancement with gadolinium was present in 92% (22/24) of children with Guillain-Barré syndrome on initial spinal magnetic resonance imaging (95% confidence interval, 0.745-0.978). This finding increased to 100% of patients, after two patients underwent repeat spinal magnetic resonance imaging that did reveal nerve root enhancement. Patterns of enhancement were variable, but involved the thoracolumbar nerve roots in all patients. Enhancement of nerve roots with gadolinium on initial spinal magnetic resonance imaging was frequently present in these children with Guillain-Barré syndrome. Spinal magnetic resonance imaging is a sensitive diagnostic test and should be considered an additional diagnostic tool in select cases. Published by Elsevier Inc.

Table 1. Patient demographics, clinical presentations, diagnostic testing, and treatments

Patient	Year	Age (yr)	Sex	Signs	Spinal MRI	Nerve Root Enhancement	Days to MRI	CSF ro (mg/dL)	NCS/EMG	Score (A-C)*	Treatment
1	2002	4	F	Gait change	C-L	Positive	6	78	N	A	Plasmapheresis
2	2004	2	M	Weakness, HR	C-L	Positive	15	597	N	B	IVIG
3	2004	3	M	Weakness, HR	C-L	Positive	5	59	Y	A	IVIG + Plasmapheresis
4	2004	11	M	Weakness, HR	C-L	Positive	26	147	N	C	IVIG + Plasmapheresis
5	2007	9	F	Facial paralysis, weakness, AR	C-L	Positive	9	45	N	A	IVIG
6	2006	3	M	Weakness, AR	C-L	Positive	14	45	N	A	None
7	2007	2	M	MFV (ataxia, ophthalmoplegia), weakness, AR	C-L	Positive	5	64	N	A	IVIG
8	2008	8	F	Weakness, HR	C-L	Positive	61	120	N	A	None
9	2008	9	M	Weakness, AR	C-L	Positive	2	101	N	B	IVIG
10	2003	15	F	Weakness, AR	C-L	Negative	1	78	Y	B	IVIG + Plasmapheresis
					C-L	Positive	12			A	
11	2005	14	M	Weakness, HR	L	Positive	7	52	Y	A	IVIG
12	2005	7	F	Weakness, AR	C-L	Positive	7	184	N	A	IVIG
13	2005	18	F	MFV (ataxia, ophthalmoplegia), weakness, AR	C-L	Positive	5	32	Y	C	IVIG + Plasmapheresis
14	2005	15	F	Weakness, AR	C-L	Positive	10	32	Y	B	IVIG + Plasmapheresis
15	2005	5	M	Weakness, AR	C-L	Positive	4	80	N	A	IVIG
16	2006	13	M	Weakness, including bulbar, HR	C-L	Negative	3	38	Y	B	IVIG + Plasmapheresis
					C-L	Positive	7			B	
17	2006	19	F	Weakness, AR	L	Positive	42	178	N	B	IVIG + Plasmapheresis
18	2006	6	F	Weakness, AR	L	Positive	14	208	N	A	IVIG
19	2006	3	M	Weakness, AR	L	Positive	32	67	N	A	IVIG
20	2007	1	M	Weakness, AR	C-L	Positive	2	150	N	B	IVIG + Plasmapheresis
21	2007	1	F	Vagal nerve and ocular involvement, weakness, AR	C-L	Positive	4	120	N	C	IVIG
22	2007	17	M	Facial weakness, bulbar involvement, weakness, AR	C-L	Positive	9	90	N	A	IVIG
23	2009	8	M	Weakness, AR	C-L	Positive	22	192	N	A	None
24	2009	4	F	Weakness, AR	C-L	Positive	6	266	N	B	IVIG

* Patients with score A were ambulatory at admission, those with score B were non-ambulatory at admission, and those with score C required mechanical ventilation at time of admission.

DIAGNOSTICO DIFERENCIAL DEL S. GUILLAIN-BARRE

Intracranial/spinal cord abnormalities
Brainstem encephalitis, meningitis carcinomatosis/ lymphomatosis, transverse myelitis, cord compression

Anterior horn cell abnormalities
Poliomyelitis, West Nile virus

Spinal nerve root abnormalities
Compression, inflammation (eg, cytomegalovirus), leptomeningeal malignancy

Peripheral nerve abnormalities
CIDP, drug-induced neuropathy, porphyria, critical illness polyneuropathy, vasculitis, diphtheria, vitamin B1 deficiency (beri-beri), heavy metal or drug intoxication, tick paralysis, metabolic disturbances (hypokalaemia, hypophosphataemia, hypermagnesaemia, hypoglycaemia)

Neuromuscular junction abnormalities
Myasthenia gravis, botulism, organophosphate poisoning

Muscular abnormalities
Critical illness polyneuromyopathy, polymyositis, dermatomyositis, acute rhabdomyolysis

CIDP=chronic inflammatory demyelinating polyneuropathy.

Van Doorn and Ruts.- Lancet Neurol 2008; 7:939-950

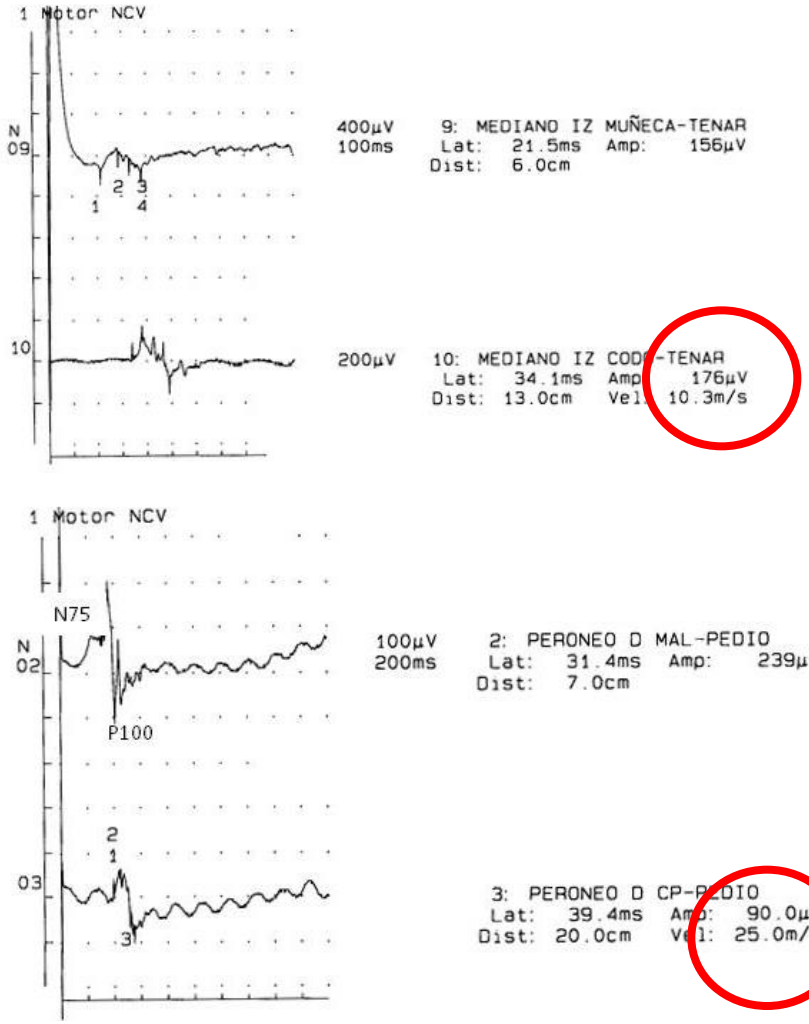
Síndrome de Guillain-Barré.

VARIANTES CLINICAS

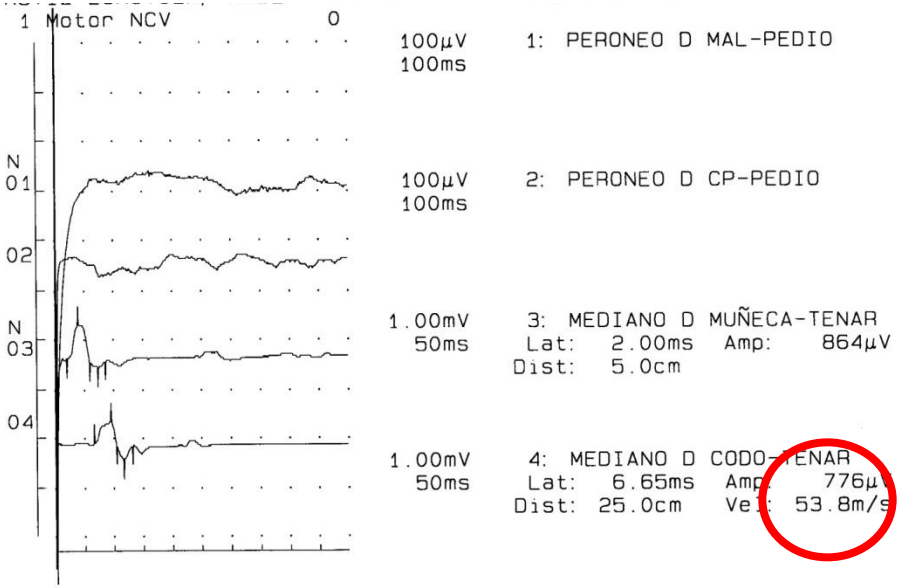
1.- **SÍNDROME G-B AGUDO DESMIELINIZANTE** (más del 85-90%).
2.- **SÍNDROME G-B AGUDO AXONAL.** Se han descrito dos tipos:
a)MOTOR Y SENSITIVO, de peor evolución que la forma desmielinizante, y
b) MOTOR (sin afectación de los nervios sensitivos).

3.- **SINDROME DE MILLER-FISHER.**
Triada **oftalmoplejia, ataxia y arreflexia**. Está desencadenado por ciertas cepas de Campylobacter jejuni que inducen la formación de Ac anti ganglióside GQ1b.

GB desmielinizante



GB axonal



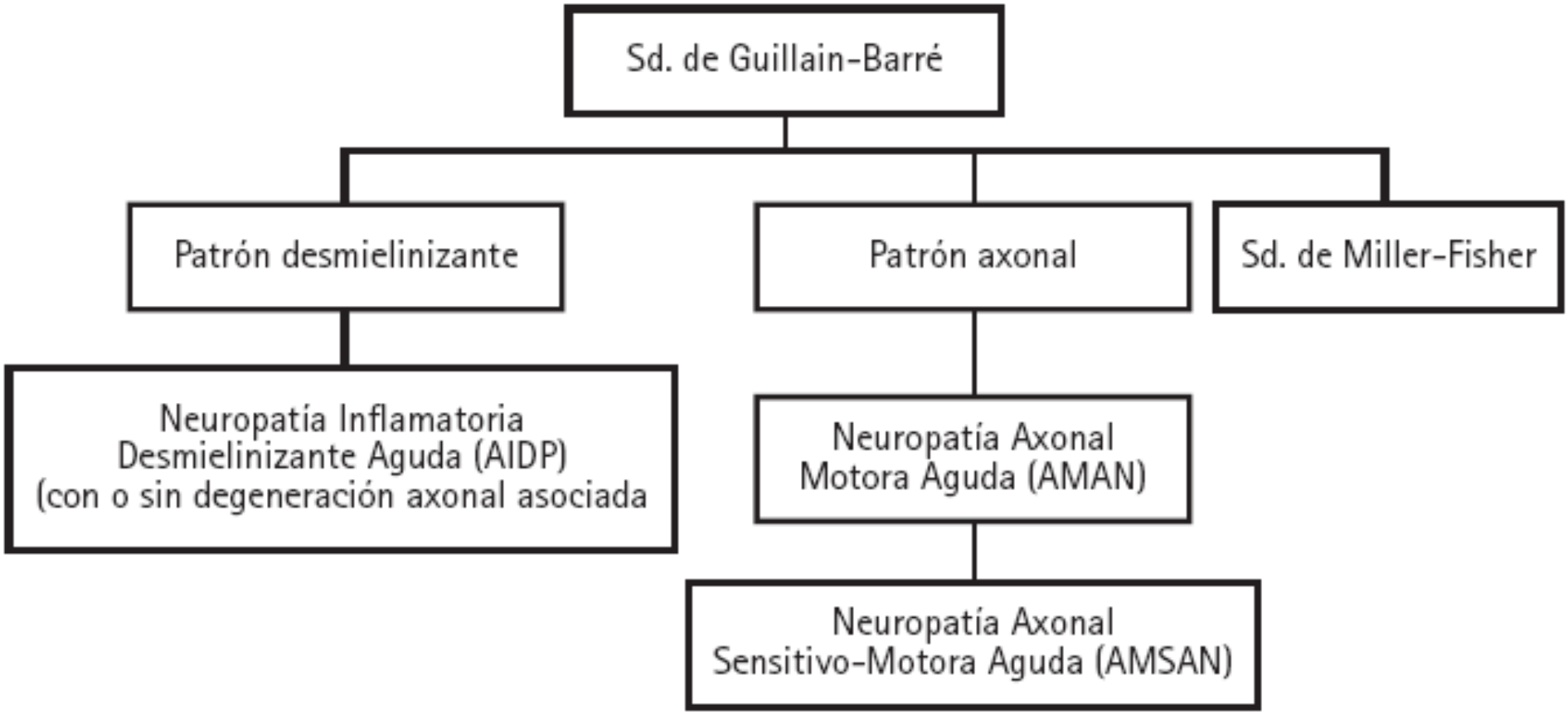
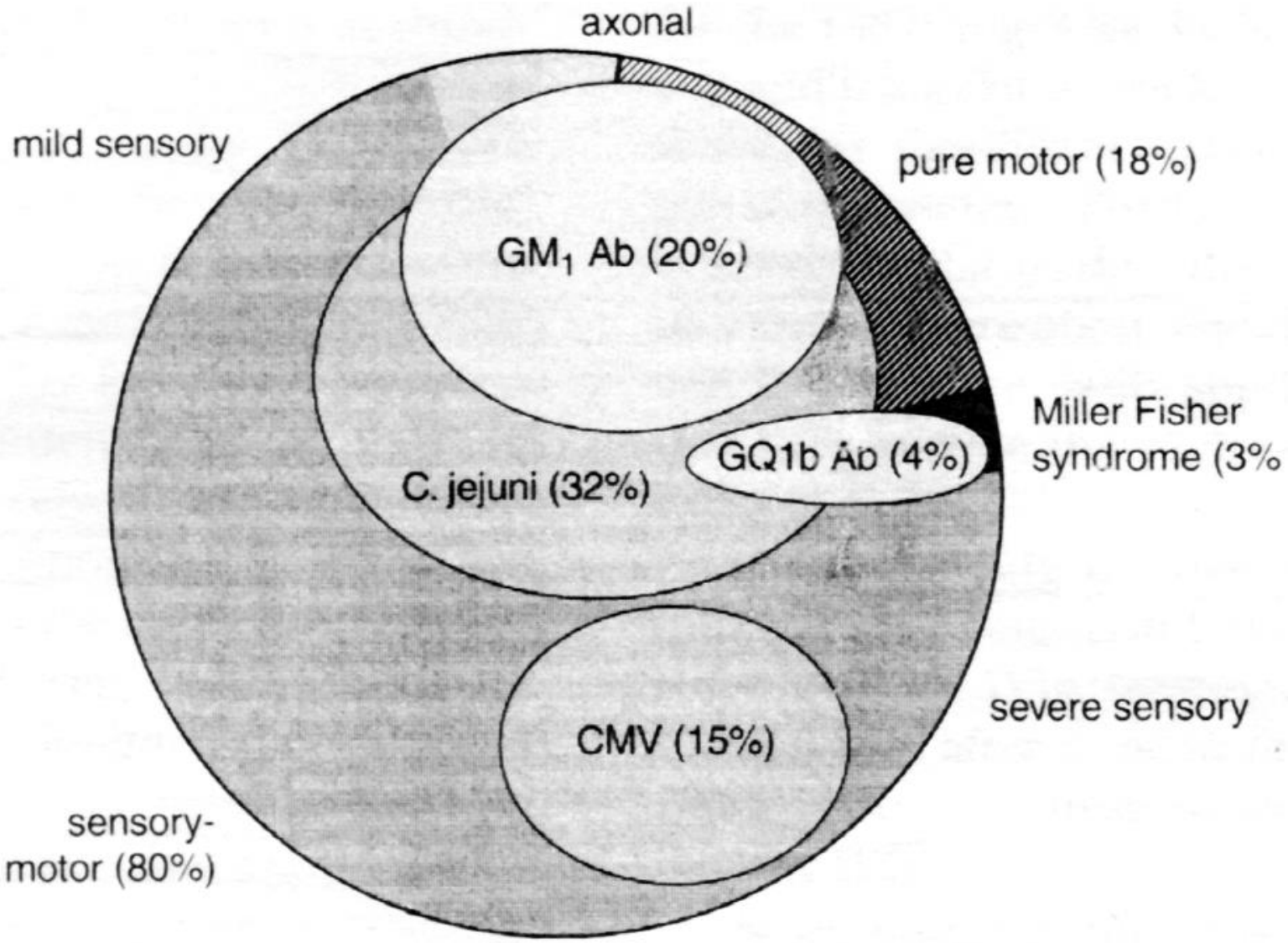

Sd. de Guillain-Barré
Patrón desmielinizante
Patrón axonal
Sd. de Miller-Fisher
Neuropatía Inflamatoria
Desmielinizante Aguda (AIDP)
(con o sin degeneración axonal asociada
Neuropatía Axonal
Motora Aguda (AMAN)
Neuropatía Axonal
Sensitivo-Motora Aguda (AMSAN)

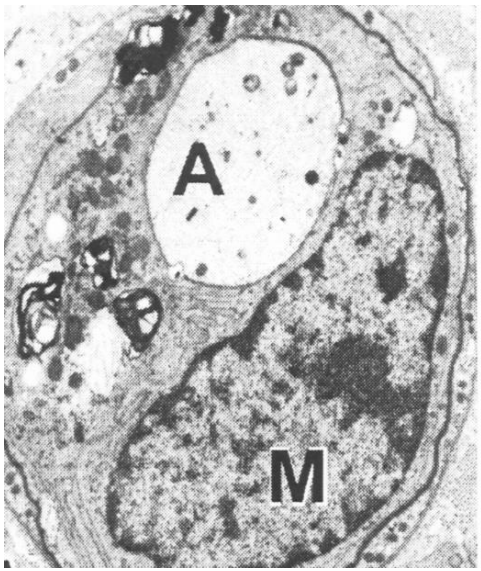
Table 1. Clinical variants of Guillain–Barré syndrome in childhood

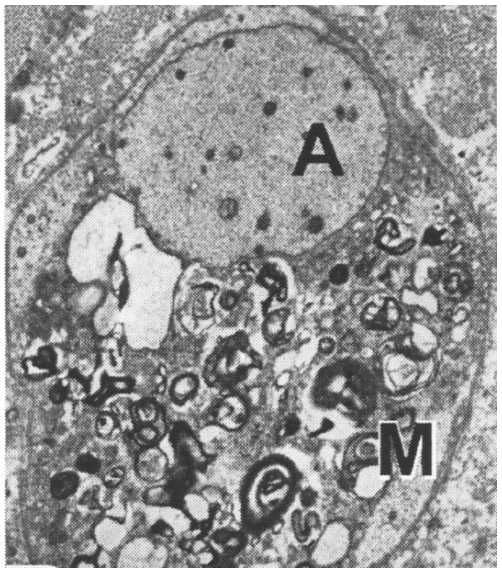
Clinical syndrome	Relative frequency	IgG antiganglioside antibody association(s)
Acute inflammatory demyelinating polyneuropathy	Common	GM1 (minority)
Acute motor axonal neuropathy	Common	GM1, GD1a
Acute motor and sensory axonal neuropathy	Uncommon	GM1, GD1a
Miller–Fisher syndrome	Uncommon	GQ1b, GT1a
Pharyngeal-cervical-brachial variant	Rare	GT1a, GQ1b, GD1a
Polyneuritis cranialis	Rare	GQ1b, GT1a
Acute (ataxic) sensory neuropathy	Very rare	GQ1b, GT1a
Acute pandysautonomia	Very rare	
Acute ophthalmoparesis	Very rare	GQ1b, GT1a

Ryan MM: Pediatric Guillain-Barré Syndrome. Curr Opin Pediatrics, 2013;25: 689-693

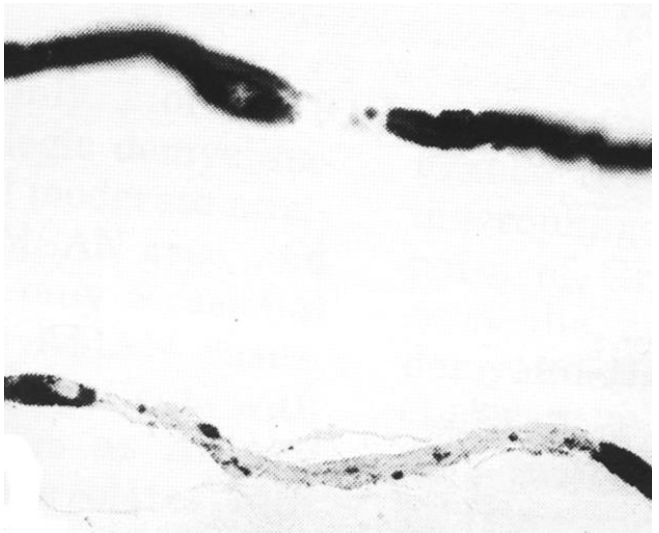

axonal
mild sensory
pure motor (18%)
GM1 Ab (20%)
Miller Fisher
syndrome (3%
GQ1b Ab (4%)
C. jejuni (32%)
severe sensory
CMV (15%)
sensory-
motor (80%)

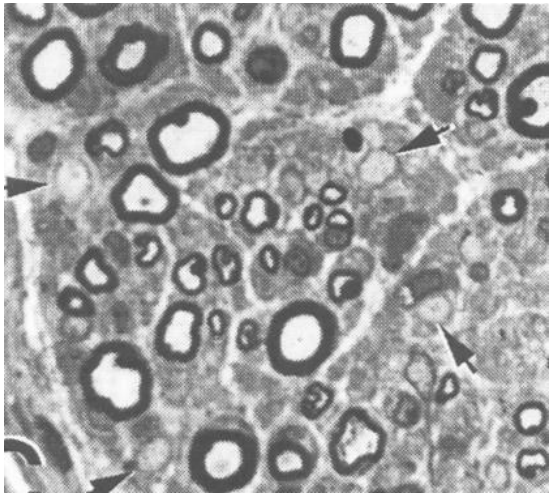
GB desmielinizante

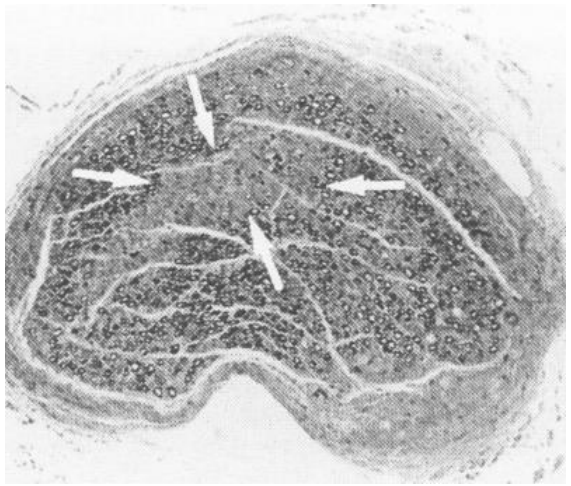




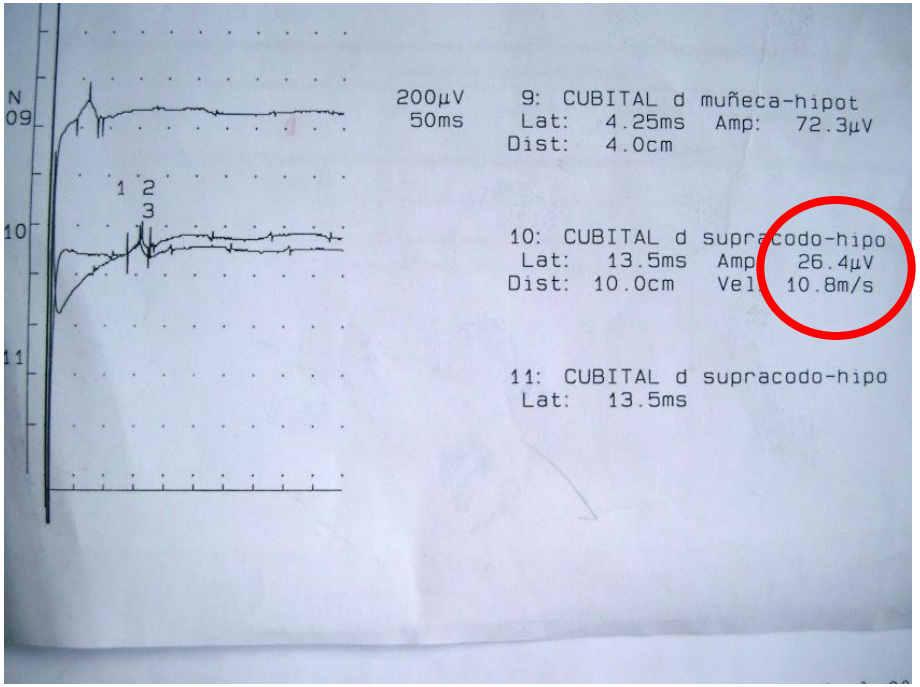
GB desmielinizante-teasing

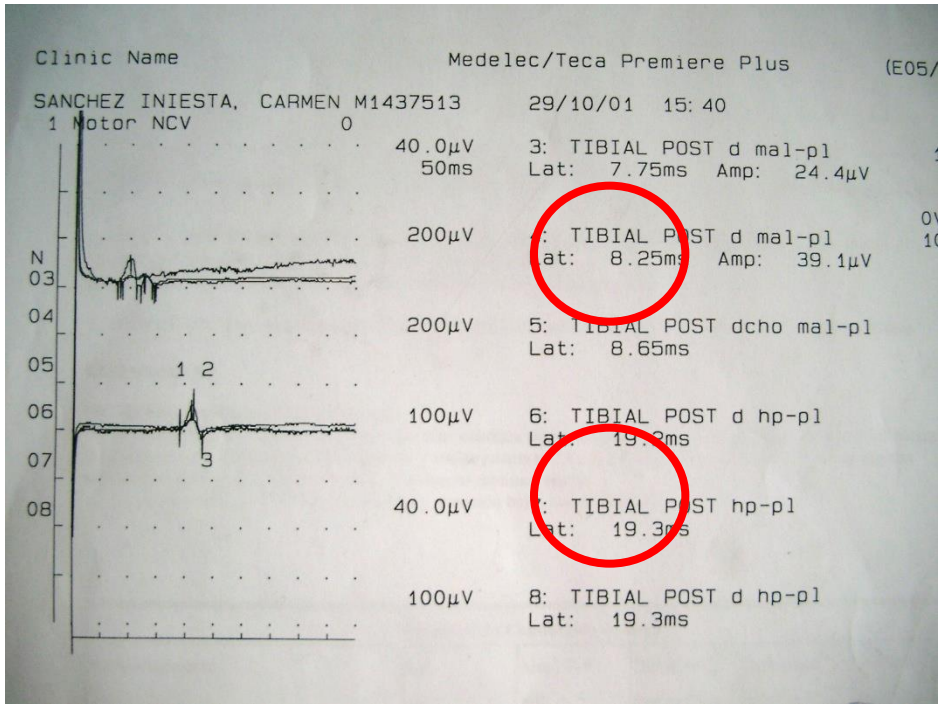




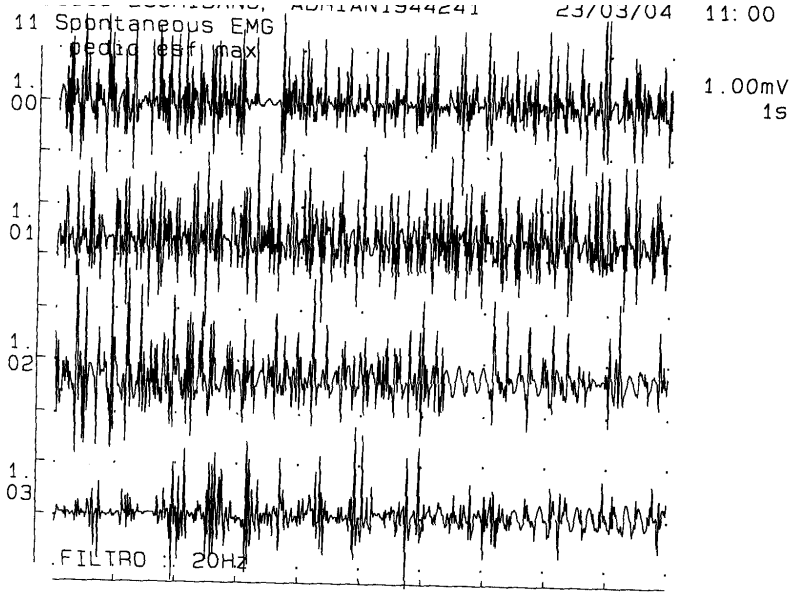


GB desmielinizante+axonal severisimo
Carmen I. 539





GB motor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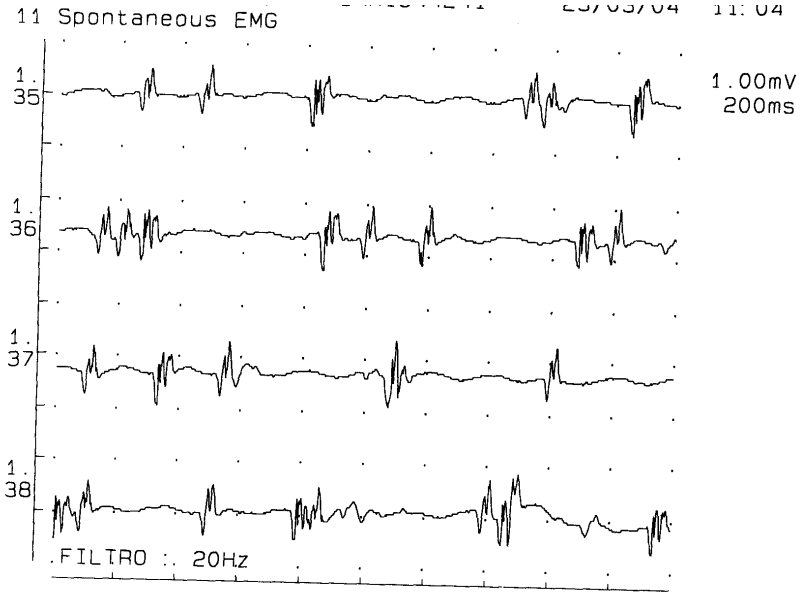
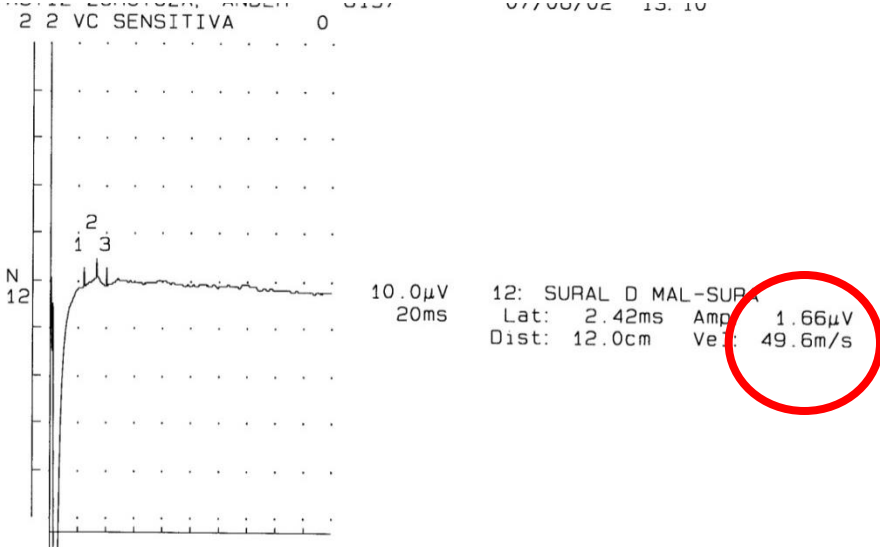
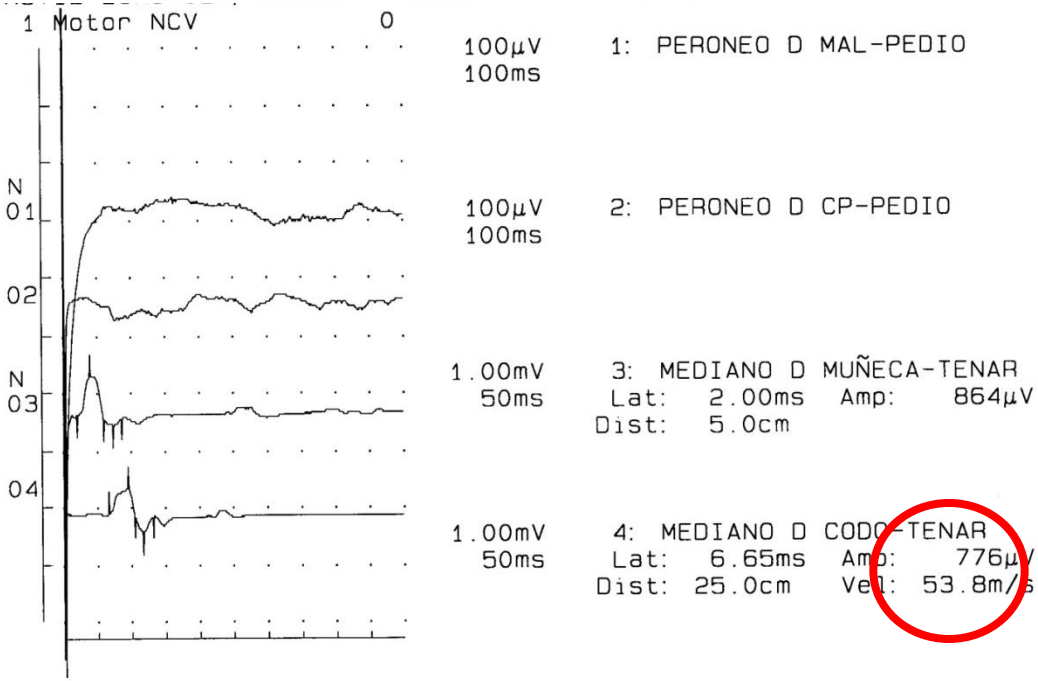
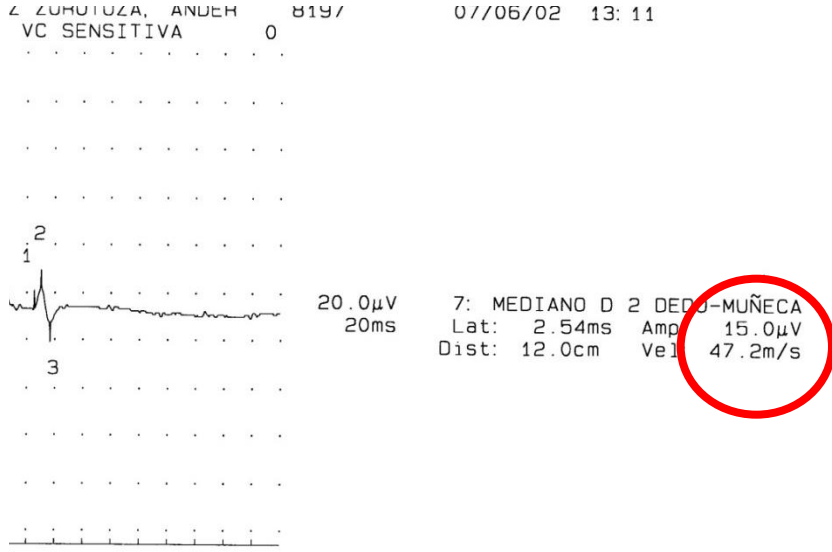
23/03/04
11: 00
1.00mV
1s
1.00
1.01
1.02
1.03
FILTRO : 20Hz


11 Spontaneous EMG
11: 04
1.00mV
200ms
1.35
1.36
1.37
1.38
FILTRO : 20Hz

GB axonal.
1863





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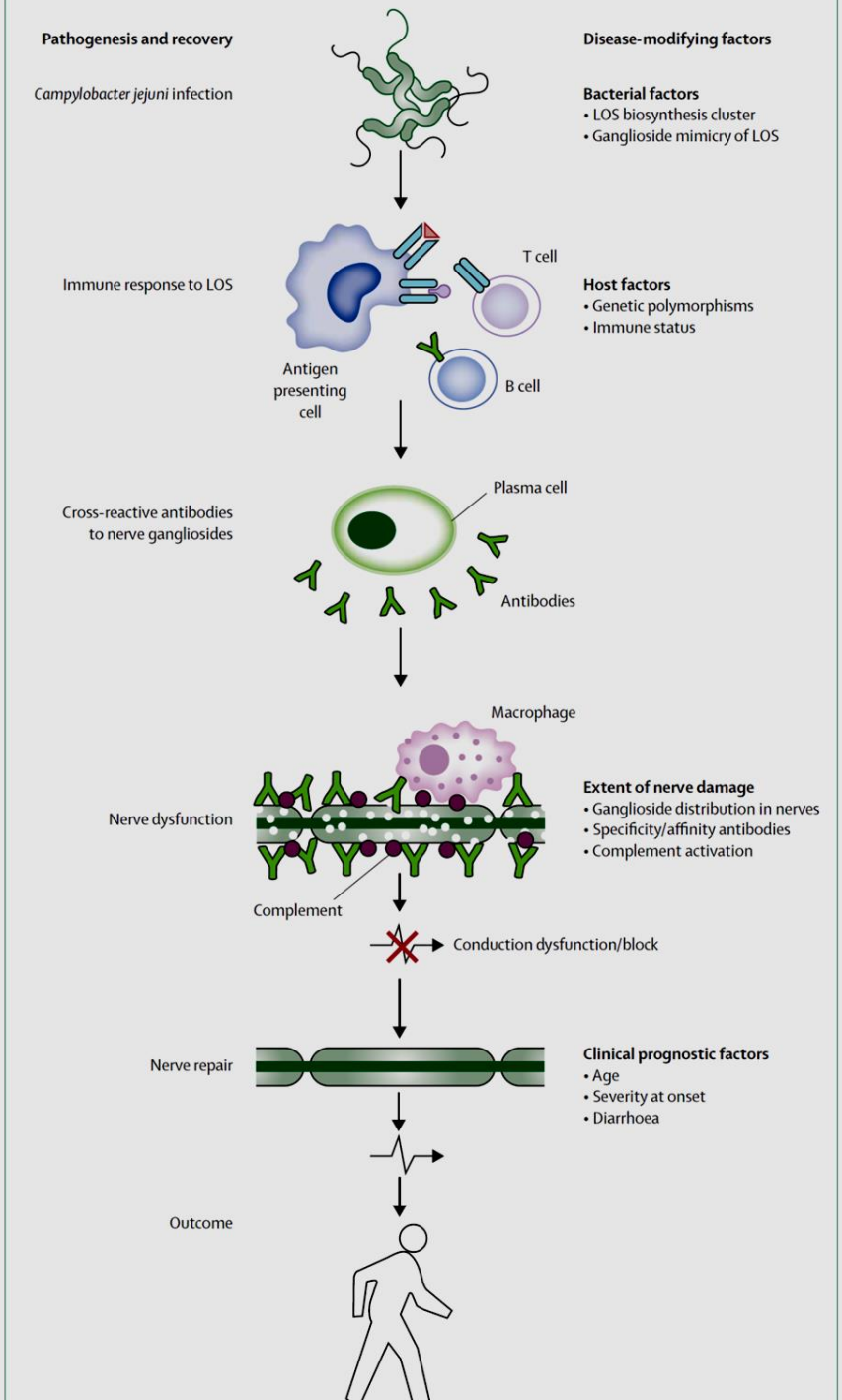


PATOGENIA

Infección (ej Campilobacter) → Respuesta inmune

Depende de:

factores bacterianos (especificidad de lipooligosacáridos LOS)
Y del huesped

Ac anti LOS reaccionan contra gangliosidos y pueden activar complemento

La gravedad depende de varios factores

ESCALA FUNCIONAL DE GRAVEDAD CLINICA

0.- sano, normal.
1.- síntomas y signos leves, pero que le permiten hacer las actividades de andar, correr aún con dificultad, actividades de vestido, comida y aseo.
2.- puede caminar más de 10 metros sin ayuda ni apoyo, pero no saltar o realizar actividades para su cuidado personal.
3.- puede caminar más de 10 metros pero con ayuda o apoyo.
4.- está confinado en cama.
5.- con ventilación asistida a tiempo total o parcial.
6.- muerte

Pascual Pascual, http://www.aeped.es/documentos/protocolos-neurologia

Plasma exchange/Sandoglobulin Guillain-Barré Syndrome Trial Group. Randomized trial of plasma exchange, intravenous inmunoglobulin and combined treatment in Guillain-Barré Syndrome. Lancet 1997;349:225-230.

Guillain Barre axonal motor

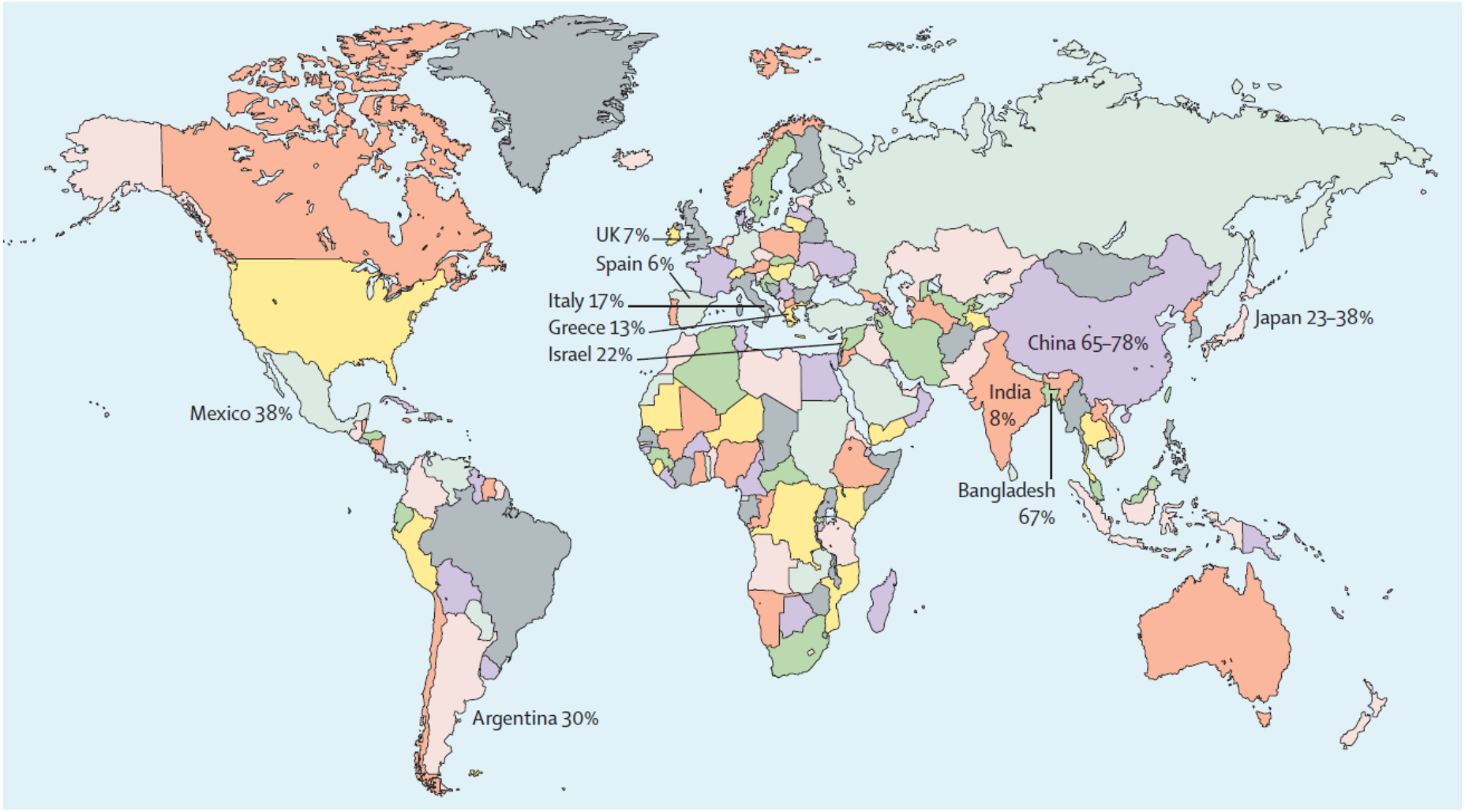
Axonal Guillain-Barré syndrome: concepts and controversies

Satoshi Kuwabara, Nobuhiro Yuki

Acute motor axonal neuropathy (AMAN) is a pure motor axonal subtype of Guillain-Barré syndrome (GBS) that was identified in the late 1990s. In Asia and Central and South America, it is the major subtype of GBS, seen in 30–65% of patients. AMAN progresses more rapidly and has an earlier peak than demyelinating GBS; tendon reflexes are relatively preserved or even exaggerated, and autonomic dysfunction is rare. One of the main causes is molecular mimicry of human gangliosides by *Campylobacter jejuni* lipo-oligosaccharides. In addition to axonal degeneration, electrophysiology shows rapidly reversible nerve conduction blockade or slowing, presumably due to pathological changes at the nodes or paranodes. Autoantibodies that bind to GM1 or GD1a gangliosides at the nodes of Ranvier activate complement and disrupt sodium-channel clusters and axoglial junctions, which leads to nerve conduction failure and muscle weakness. Improved understanding of the disease mechanism and pathophysiology might lead to new treatment options and improve the outlook for patients with AMAN.

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Guillain Barre axonal motor

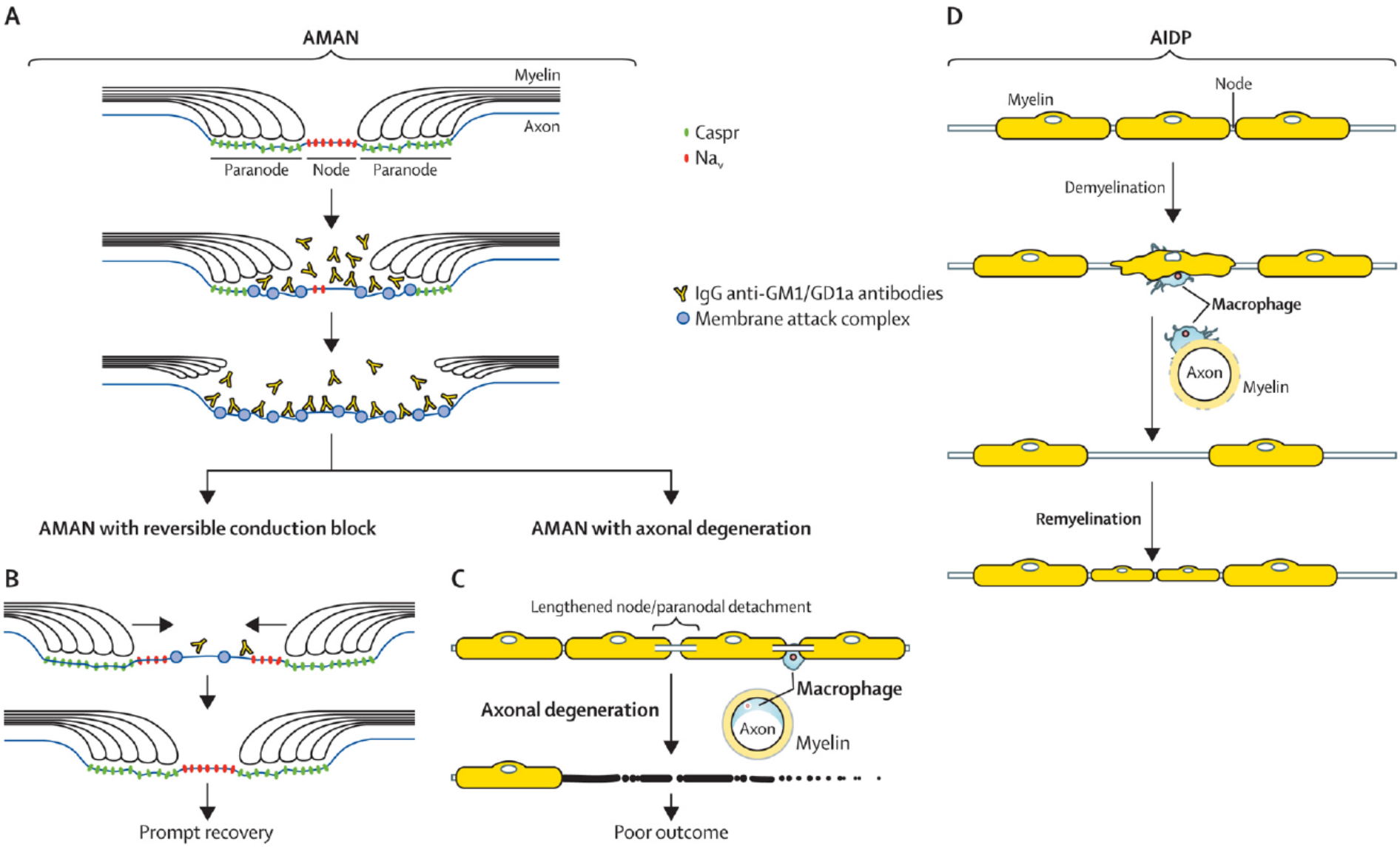


Frecuencias de Polineuropatia axonal aguda motora en pacientes con Sindrome G-B

Ho et al. Brain 1995; 118: 597–605.

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Guillain Barre axonal motor.- Inmunopatogenia



Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Guillain Barre axonal motor (AMAN).- Criterios electrofisiológicos

	Ho and colleagues[3]	Hadden and colleagues[17]
Acute inflammatory demyelinating polyneuropathy	Must have at least one of the following in two or more nerves: conduction velocity <90% LLN or <85% if distal CMAP <50% LLN; distal latency >110% ULN or >120% if distal CMAP is <LLN; unequivocal temporal dispersion; and F-wave latency >120% ULN	Must have at least one of the following in two or more nerves: conduction velocity <90% LLN or <85% if distal CMAP <50% LLN; distal latency >110% ULN or >120% if distal CMAP <LLN; ratio of proximal-to-distal amplitude <0·5 and distal CMAP ≥20% LLN; and F-wave latency >120% ULN
Acute motor axonal neuropathy	No evidence of demyelination and distal CMAP <80% of LLN	None of the above (or in only one nerve if distal CMAP <10% of LLN) and distal CMAP <80% in two nerves

LLN=lower limit of normal. ULN=upper limit of normal. CMAP=compound-muscle action potential.

Table 1: Electrodiagnostic criteria for acute inflammatory demyelinating polyneuropathy and acute motor axonal neuropathy

Ho et al. Brain 1995; 118: 597–605.

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Guillain Barre axonal Motor .- clinica

La infección previa por Campylobacter Jejuni es la más frecuente

	Acute motor axonal neuropathy	Acute inflammatory demyelinating polyneuropathy
Preceding infection	*Campylobacter jejuni*	Cytomegalovirus, Epstein-Barr virus
Frequency	Europe and North America <10%; Mexico 30%; Israel 22%; China 65%; Japan 38%	Europe and North America 90%; Mexico 30%; Israel 63%; China 20%; Japan 40%
Epidemics	Children (in China and Mexico)	None
Cranial nerve palsy	Rare (<20%)	Frequent (60%)
Sensory loss	Usually none (<10%)	Frequent (70%)
Pain	Usually none	Frequent (up to 66%)
Autonomic involvement	Rare	Frequent
Tendon reflex	Usually absent (preserved or exaggerated in 20%)	Absent (preserved or exaggerated in 5%)
Recovery	Two patterns (rapid and slow)	Relatively uniform
Electrophysiology	Axonal degeneration Reversible conduction blockade or slowing	Demyelination
Target molecule	Gangliosides (eg, GM1 and GD1a)	Unknown

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Guillain Barre axonal motor (AMAN)- Evolución

Es parecida a la de la forma desmielinizante, pero hay dos evoluciones diferentes

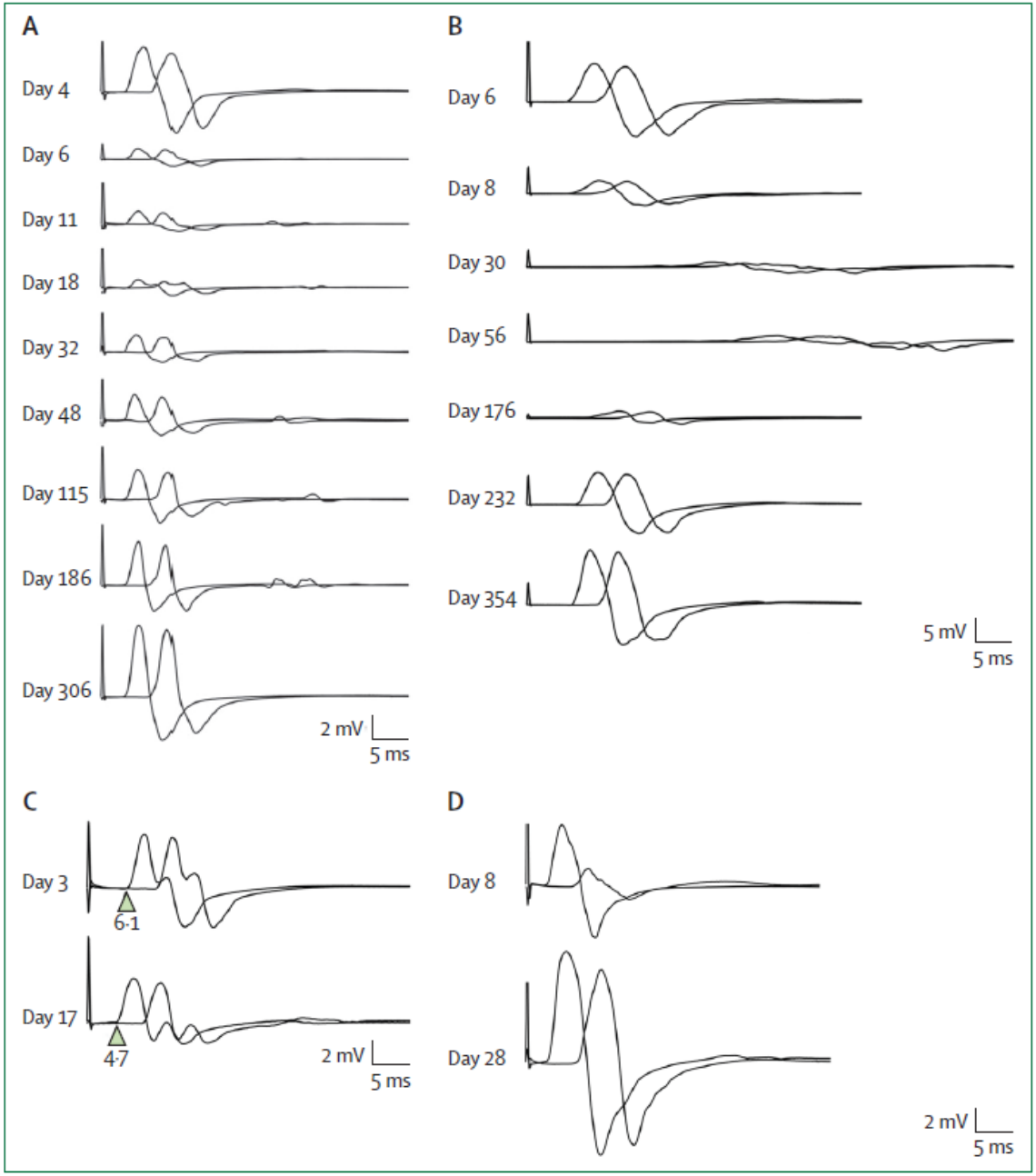
1.- Recuperación muy rápida, en días (bloqueo conducción)
2.- Otros se recuperan muy lentamente y con secuelas. (degeneración axonal proximal , en las raices nerviosas).

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Guillain Barre axonal.- Neurofisiologia. Evolución

Axonal motor

Desmielinizante



Axonal motor

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

Síndr. de Guillain-Barré.

TRATAMIENTO

- **Ingreso en un centro con UCI y experiencia en el manejo de esta patología.**
- **Monitorización pulmonar**
- **Disfunción autonómica (FC, TA, ileo)**
- **Rehabilitación temprana**

- **Plasmaféresis ó inmunoglobulinas IV.**

Ambas han demostrado similar utilidad.

Pascual Pascual http://www.aeped.es/documentos/protocolos-neurologia

ESCALA FUNCIONAL DE GRAVEDAD CLINICA (GBS disability Score)

0.- sano, normal.
1.- síntomas y signos leves, pero que le permiten hacer las actividades de andar, correr aún con dificultad, actividades de vestido, comida y aseo.
2.- puede caminar más de 10 metros sin ayuda ni apoyo, pero no saltar o realizar actividades para su cuidado personal.
3.- puede caminar más de 10 metros pero con ayuda o apoyo.
4.- está confinado en cama.
5.- con ventilación asistida a tiempo total o parcial.
6.- muerte

Plasma exchange/Sandoglobulin Guillain-Barré Syndrome Trial Group. Lancet 1997;349:225-230.

Pascual Pascual http://www.aeped.es/documentos/protocolos-neurologia

Síndr. de Guillain-Barré.

A QUÉ PACIENTES HAY QUE TRATAR

Debe llegarse al diagnóstico y tomar la decisión de tratar lo antes posible.

Se pueden considerar tres tipos de evolución precoz de la enfermedad:

1.- Empeoramiento en la situación funcional:

En el estadio 3 →tratarla (IgG IV o plasmaféresis) lo antes posible.

2.- Curso estable, tras el empeoramiento inicial.

Previsible evolución benigna.

La actitud será de vigilancia. Solo se tratará en caso de retroceso funcional.

3.- Mejora progresiva. No tratar con IgG o plasmaféresis.

Tras comenzar el tratamiento (IgG IV ó plasmaféresis) todavía 1/4 a 1/3 de los casos pueden mostrar deterioro durante unos días. No es apropiado, por tanto, cambiar de uno a otro tratamiento.

Mientras no haya otra información, conviene completar el tratamiento que se haya iniciado, sin cambiar a otro.

Pascual Pascual http://www.aeped.es/documentos/protocolos-neurologia

Síndr. de Guillain-Barré.

INMUNOGLOBULINA INTRAVENOSA.

Comenzar el tratamiento lo antes posible.
0,4 gr/k de peso y día durante 5 días.
En niños el resultado es el mismo con la misma dosis total en 2 días (1 gr/k/dia)
Recaídas en un 5% , se tratan con una nueva dosis de 0,4 gr/K en un día.
La tasa de recaídas es similar tras plasmaféresis

PLASMAFERESIS.

Debe realizarse cuanto antes, preferiblemente en la primera semana, aunque puede llegar a ser útil incluso en el primer mes.
Minimo nº de recambios: dos sesiones.
En los casos leves (estadio funcional de 2 ó 3) basta con dos sesiones.
Los casos moderados (estadio 4) evolucionan mejor con 4 sesiones que con dos.
Los casos graves (estadio 5) requieren cuatro recambios, y su evolución no mejora aumentando a seis.
Los recambios son de unos 40 cc/k en cada uno, y se hacen a días alternos.
Las recaídas (empeoramiento 1-2 semanas tras la mejoría inicial) se pueden tratar con nuevos recambios plasmáticos, o bien con IgG IV.

CORTICOIDES.- No mejoran la evolucion (oral o IV)

G-B
Evolución

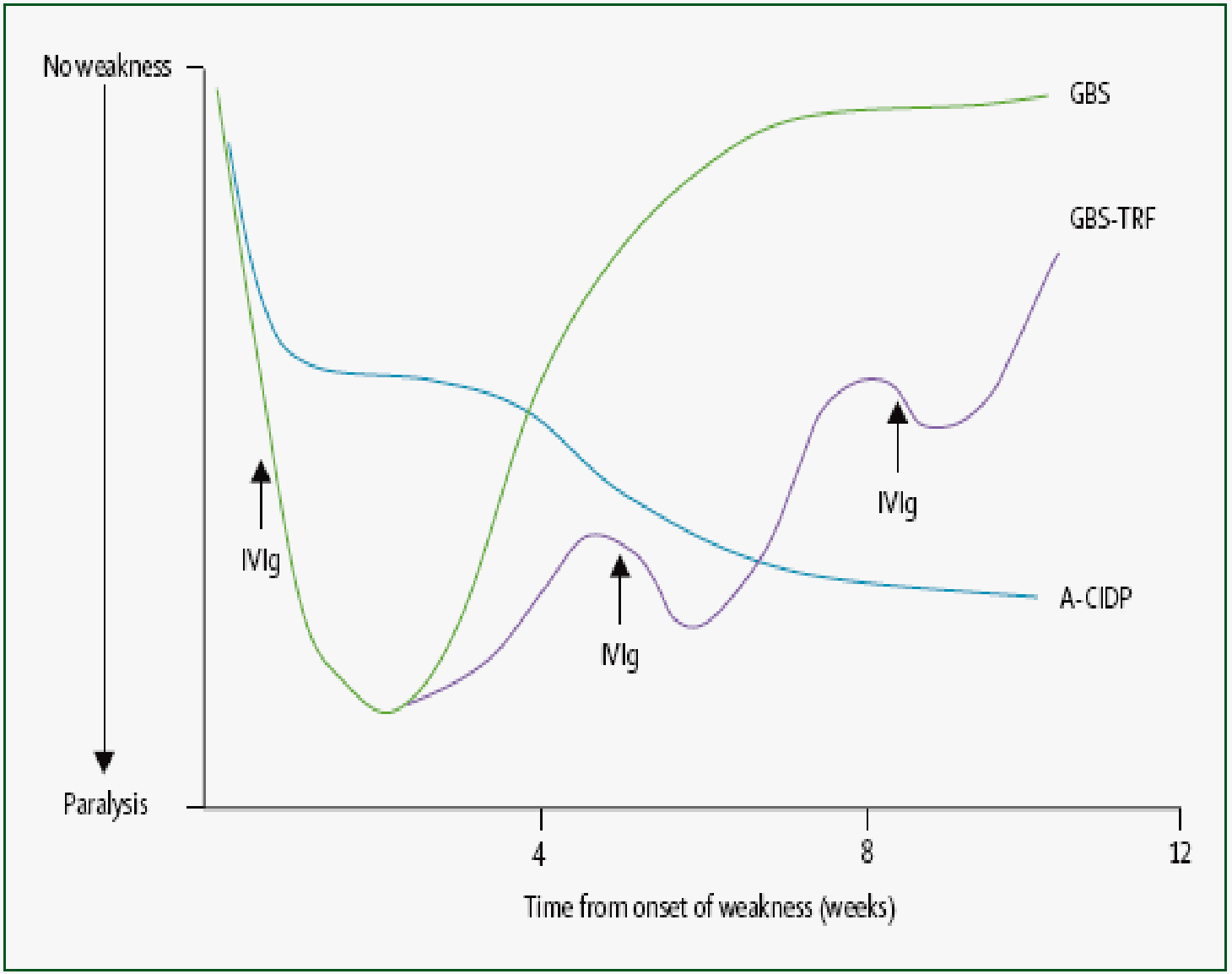


Figure 3: GBS, treatment-related fluctuations (TRF), and acute-onset CIDP (A-CIDP)
IVIg=treatment with a course of IVIg (2 g/kg bodyweight) over 2–5 days.

Van Doorn and Ruts.- Lancet Neurol 2008; 7:939-950

Guillain Barre axonal.- Tratamiento

- No está todavia bien determinada la utilidad de IgG IV y plasmaféresis en AMAN .

- Estudios preliminares parecen indicar mejor respuesta a la IgG IV

- Posibilidad de que los inhibidores del complemento puedan ser útiles para evitar las formas graves de AMAN

Kuwabara et Yuki. Lancet Neurol 2013; 12: 1180–88

A clinical prognostic scoring system for Guillain-Barré syndrome

Lancet Neurol 2007; 6: 589–94

Rinske van Koningsveld, Ewout W Steyerberg, Richard A C Hughes, Anthony V Swan, Pieter A van Doorn, Bart C Jacobs

Summary

Background Guillain-Barré syndrome (GBS) is an acute post-infectious immune-mediated peripheral neuropathy with a highly variable clinical course and outcome. We aimed to develop and validate a scoring system based on clinical characteristics in the acute phase of GBS to predict outcome at 6 months.

Methods We studied patients with GBS who were unable to walk independently. A derivation set included 388 patients from two randomised controlled trials and one pilot study. Potential predictors were assessed for their association with the inability to walk independently at 6 months. A simple clinical scoring system was developed on the basis of regression coefficients of predictors in a multivariable logistic regression model. Model performance was quantified with respect to discrimination (area under receiver operating characteristics curve, AUC) and calibration (graphically). We validated our scoring system in a set of 374 patients from another randomised trial.

Findings We included three variables that were predictive of poor outcome at 6 months in our model: age, preceding diarrhoea, and GBS disability score at 2 weeks after entry. Scores ranged from 1 to 7, with three categories for age, two for diarrhoea, and five for GBS disability score at 2 weeks. Predictions corresponding to these prognostic scores ranged from 1% to 83% for the inability to walk independently at 6 months. Predictions agreed well with observed outcome frequencies (adequate calibration) and showed a very good discriminative ability (AUC 0·85) in both data sets.

Interpretation A simple scoring system for patients with GBS, based on three clinical characteristics, accurately predicts outcome at 6 months. The system could be used to counsel individual patients and identify high-risk groups to guide future trials.

Lancet Neurol 2007; 6: 589–94

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Panel: Clinical scores for GBS

GBS disability score[15]

0 A healthy state
1 Minor symptoms and capable of running
2 Able to walk 10 m or more without assistance but unable to run
3 Able to walk 10 m across an open space with help
4 Bedridden or chairbound
5 Requiring assisted ventilation for at least part of the day
6 Dead

Medical Research Council (MRC) sum score[16]

Sum of MRC scores of six muscle groups, including shoulder abductors, elbow flexors, wrist extensors, hip flexors, knee extensors, and foot dorsiflexors on both sides, ranging from 60 (normal) to 0 (quadriplegic). The MRC score of an individual muscle group ranges from 0 to 5:

0 No visible contraction
1 Visible contraction without movement of the limb
2 Active movement of the limb, but not against gravity
3 Active movement against gravity over (almost) the full range
4 Active movement against gravity and resistance
5 Normal power

	Number of patients	Number of patients with poor outcome	Odds ratio (95% CI)	p value
Demographic features				
Total	388	71 (18%)		
Age (years)				
≤40	137	13 (9%)	1	0·001
40–60	111	19 (17%)	2·0 (0·9–4·2)	
>60	140	39 (28%)	3·7 (1·9–7·3)	
Sex (male)	213	41 (19%)	1·6 (0·7–1·9)	0·6
Infections and serology				
Symptoms preceding infection*				
Diarrhoea	88	27 (31%)	2·6 (1·5–4·5)	0·001
Upper respiratory tract infection	143	17 (12%)	0·5 (0·3–0·9)	0·01
Infection serology				
Campylobacter jejuni	105	30 (29%)	2·7 (1·5–4·7)	0·001
Cytomegalovirus	42	7 (17%)	0·9 (0·4–2·2)	0·9
Epstein-Barr virus	41	9 (22%)	1·4 (0·6–3·1)	0·4
Mycoplasma pneumoniae	9	1 (5%)	0·3 (0·03–1·9)	0·2
Anti-ganglioside antibodies				
GM1	74	18 (24%)	1·7 (0·9–3·2)	0·1
GD1a	20	4 (20%)	1·2 (0·4–3·7)	0·8
Neurological deficits				
GBS disability score at entry				
3	91	7 (8%)	1	0·003
4	261	52 (20%)	3·0 (1·3–6·8)	
5	36	12 (33%)	6·0 (2·1–17)	
GBS disability score at 2 weeks				
0 or 1	15	0 (0%)	0	<0·0001
2	84	2 (2%)	1	
3	65	2 (3%)	1·3 (0·2–9·5)	
4	144	30 (21%)	11 (2·5–46)	
5	80	37 (46%)	35 (8·1–153)	
MRC sum score at entry				
60–51	47	0 (0%)	0	<0·0001
50–41	179	20 (11%)	1	
40–31	79	22 (28%)	3·1 (1·6–6·0)	
30–21	50	17 (34%)	4·1 (1·9–8·6)	
20–0	32	12 (38%)	4·8 (2·0–11)	
Weakness before entry <4 days	191	49 (26%)	2·7 (1·6–4·7)	<0·0001
Cranial nerve involvement	215	40 (19%)	1·1 (0·6–1·8)	0·7
Sensory deficits	315	61 (19%)	1·5 (0·7–3·1)	0·3

MRC=Medical Research Council. *Symptoms of an infection in the 4 weeks preceding the onset of weakness.

Table 1: **Characteristics of the derivation set of 388 patients with GBS in relation to poor outcome at 6 months (inability to walk independently)**

Lancet Neurol 2007; 6: 589–94

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	Categories	Score
Age at onset (years)	>60	1
	41–60	0·5
	≤40	0
Diarrhoea (≤4 weeks)	Absence	0
	Presence	1
GBS disability score (at 2 weeks after entry)	0 or 1	1
	2	2
	3	3
	4	4
	5	5
Erasmus GBS outcome score		1–7

: The Erasmus GBS outcome score

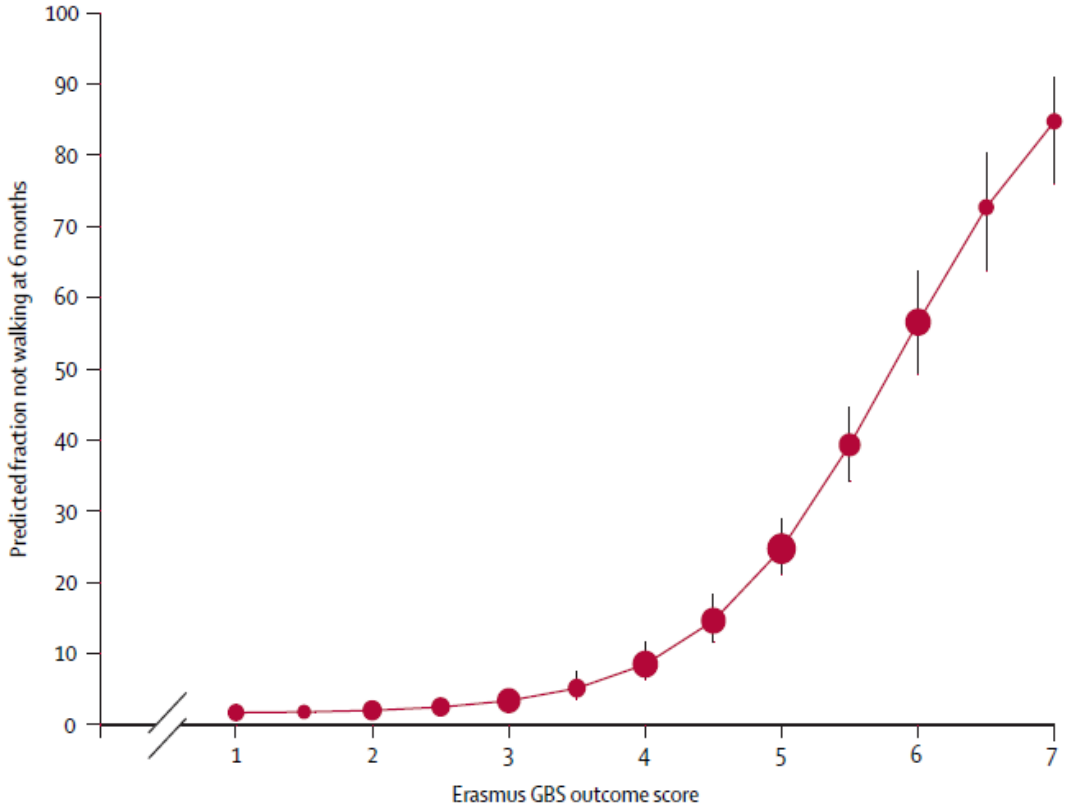

Figure: Predicted fraction of patients unable to walk independently at 6 months after randomisation on the basis of the Erasmus GBS outcome score (n=762)

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journal homepage: www.elsevier.com/locate/jns

Residual disability 10 years after falling ill in Guillain–Barré syndrome: A prospective follow-up study

Anette Forsberg [a,b,*], Rayomand Press [a,c], Lotta Widén Holmqvist [a,d]

A B S T R A C T

Objective: To describe residual disability 10 years after onset of Guillain–Barré syndrome (GBS) and longitudinal changes from 2 weeks after onset until 10 years afterwards. The Erasmus GBS Outcome score (EGOS) was applied for predicting prognosis at 2 and 10 years.

Methods: Twenty-nine patients, mean age at onset 49 years, were followed prospectively from 2 weeks to 10 years after GBS onset. Measures included; GBS disability score, EGOS, Barthel Index, Frenchay Activity Index, Sickness Impact Profile (SIP), Overall Neuropathy Limitations Scale (ONLS), Walk-12, and Fatigue Severity Scale.

Results: At 10 years, the facial paralysis found in 5 participants at 2 years was still present, 11 participants (38%) experienced paresthesia, 6 (21%) had limitations in their arms, and 15 (52%) had limitations in walking. Decreased health-related quality of life on comparison to the general population was seen in the physical dimension of SIP at 10 years. The median EGOS at 2 weeks was 4.5, which correlated highly only with the Barthel Index at 2 years and the ONLS arm scale at 10 years.

Conclusion: The residual disabilities at 1–2 years comprised mainly of reduced walking ability, and are still persistent 10 years after GBS onset. For some individuals, facial paralysis caused major disability. The EGOS only partly predicted residual disability at 2 and 10 years after onset.

GBS disability score at 2 weeks, 2 years, and 10 years after onset of GBS, pr
medians (inter-quartile range) and numbers (n = 29). The arrows show the change of scores for four patients showing some recovery during the time from 2 to 10 years after onset.

Median (IQR)	2 weeks 4 (2.5 – 5)	2 years 1 (1 – 1)	10 years 1 (0 – 1)
0 = healthy	0	10	10
1 = minor symptoms or signs, able to run	0	14	15
2 = able to walk more than 10 m without assistance but unable to run	7	1	1
3 = able to walk more than 10 m with assistance	4	1	2
4 = requires wheel-chair	10	3	1
5 = requiring assisted ventilation for at least part of the day	8	0	0
6 = dead due to Guillain–Barré syndrome	0	0	0

Treatment for Fisher syndrome, Bickerstaff's brainstem encephalitis and related disorders (Review)

Overell JR, Hsieh ST, Odaka M, Yuki N, Willison HJ

The Cochrane Library 2007, Issue 3
http://www.thecochranelibrary.com

CONCLUSION:

Despite the absence of evidence from randomised trials, the review authors consider it **clinically logical to extrapolate GBS data to the population of FS patients with complicated disease** (profound ataxia, severe bulbar palsy, or those with respiratory or limb weakness in GBS overlap syndromes).

Patients with less severe syndromes are likely to improve completely with a conservative approach.

Whether the speed of this recovery can be hastened with currently available therapies is unknown.

Polineuritis crónica inflamatoria desmielinizante

- Enfermedad desmielinizante, autoinmune.
- Rara, prevalencia, en todas las edades, de 1 a 1,9/100.000.
- En niños es menos frecuente que en adultos, prevalencia de 0,48/100.000.

- Importante pensar en este cuadro, puesto que, tiene tratamiento.

Pascual Pascual.- Rev Neurol. 2002;35:269-276

Polineuritis crónica inflamatoria desmielinizante

Criterios diagnósticos :

1. Polirradiculoneuropatía o polineuropatía simétrica, que afecta a fibras motoras y sensitivas, más distal que proximal.

   Debilidad, alteración sensitiva y parestesias que pueden comenzar de forma aguda, como un SGB, o más lenta.

2. **El curso es progresivo, continuo o en brotes**. La afectación máxima se alcanza **después de ocho semanas** tras el inicio, a diferencia del SGB;

3. Proteínas en el LCR elevadas (de 1,5 a 4 veces el valor normal), y no suele haber más de 5 células/mL;

4. La VCM y VCS es menor del 80% del valor mínimo normal y otros signos desmielinización (lat distal aumentada, bloqueos)

5. La plasmaféresis y corticosteroides mejoran claramente el cuadro.
6. Desmielinización en el nervio periférico y en las raíces. y
7. No hay signos de otra causa de la polineuropatía.

Pascual Pascual.- Rev Neurol. 2002;35:269-276

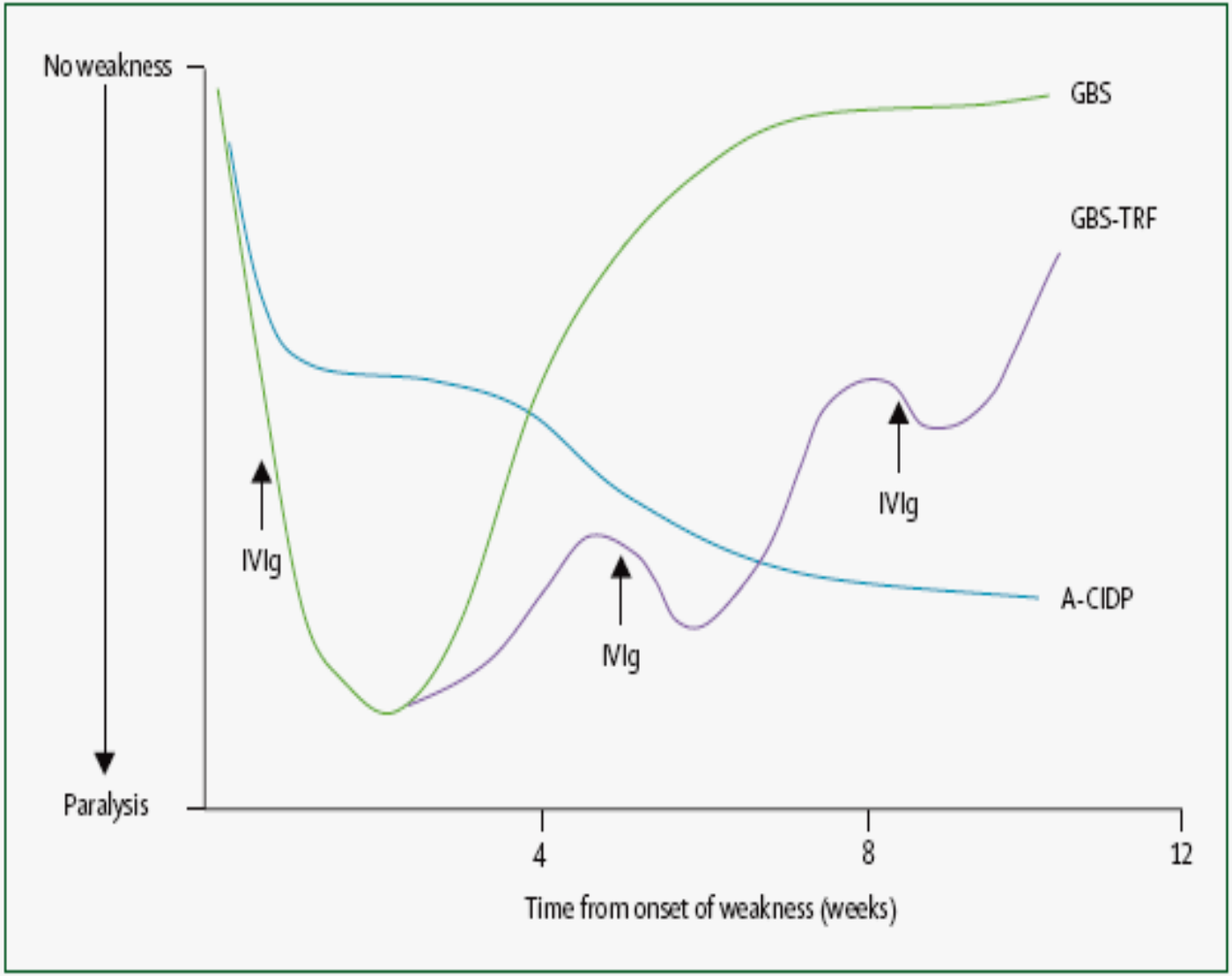
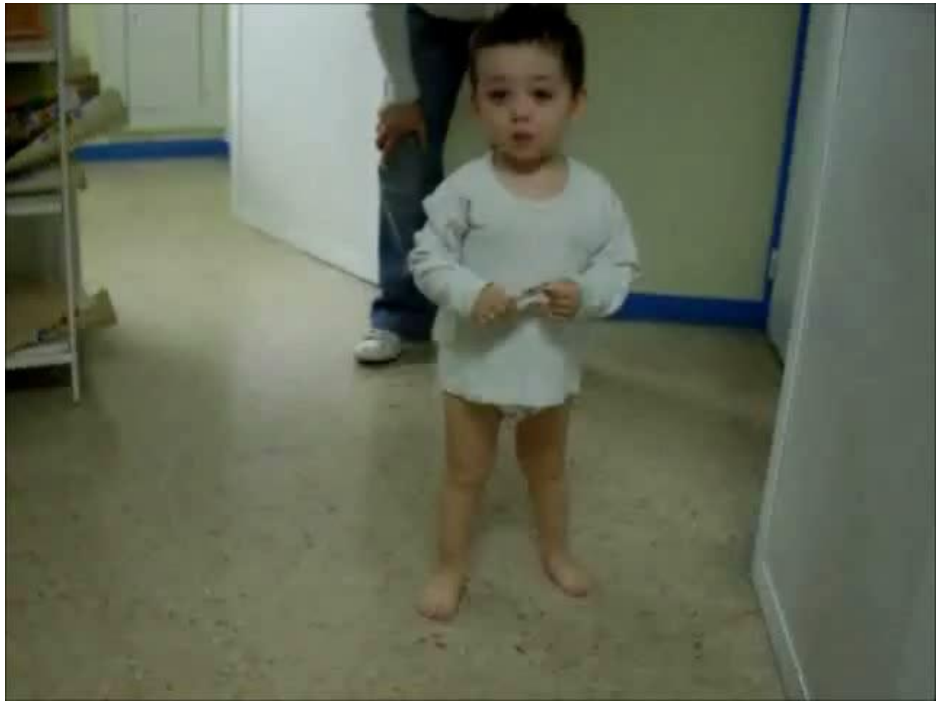


Figure 3: GBS, treatment-related fluctuations (TRF), and acute-onset CIDP (A-CIDP)
IVIg=treatment with a course of IVIg (2 g/kg bodyweight) over 2–5 days.

Van Doorn and Ruts.- Lancet Neurol 2008; 7:939-950

Polineuritis cronica inflam desmielinizante. Mario

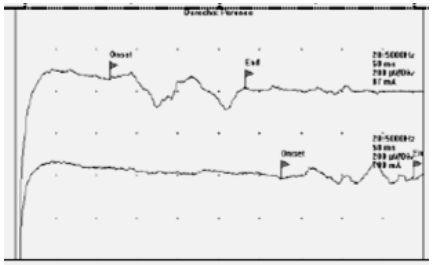
Antes de tratamiento, 2010



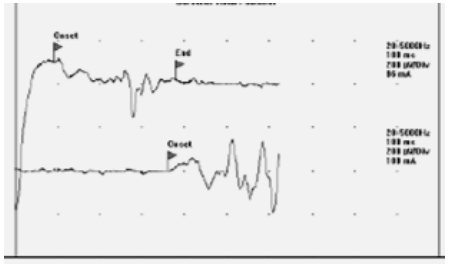
Con IgG IV + deflazacort, 2013

Mario PCID marzo 2010-Pre-tratamiento

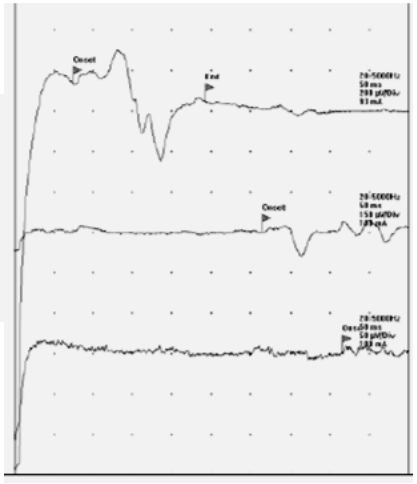
Velocidad de Conducción Nerviosa Motora						
Derecho: Peroneo	**Comienzo (ms)**	**Duración (ms)**	**Amplitud (mV)**	**Area (µVs)**	**Distancia (cm)**	**Velocidad (m/s)**
1.Maleolo- Pedio	11,6	16,6	0,2	0,8	5,0	
2.Cabeza Peronéo	32,7	16,0	0,1	0,3	15,0	7,1



Velocidad de Conducción Nerviosa Motora						
Derecho: Tibial Posterior	**Comienzo (ms)**	**Duración (ms)**	**Amplitud (mV)**	**Area (µVs)**	**Distancia (cm)**	**Velocidad (m/s)**
1.Maleolo- Planta	9,2	28,8	0,3	1,1	8,0	
2.Hueco Popliteo	36,1				14,0	5,2

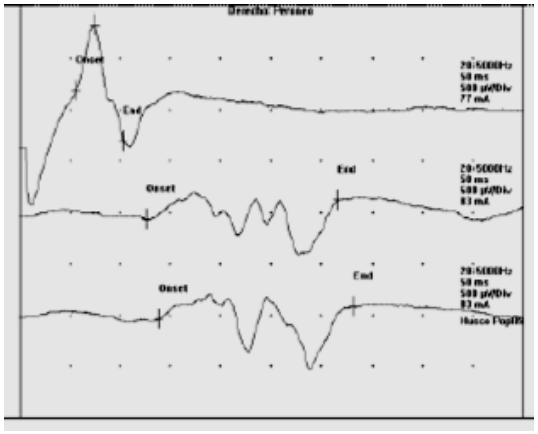


Velocidad de Conducción Nerviosa Motora						
Derecho: Mediano	**Comienzo (ms)**	**Duración (ms)**	**Amplitud (mV)**	**Area (µVs)**	**Distancia (cm)**	**Velocidad (m/s)**
1.Muñeca-Tenar	7,5	16,8	0,6	2,2	6,0	
2.Codo-Tenar	31,6		0,1		11,0	4,6
3.Axila-tenar	41,6		0,0		11,0	11,0

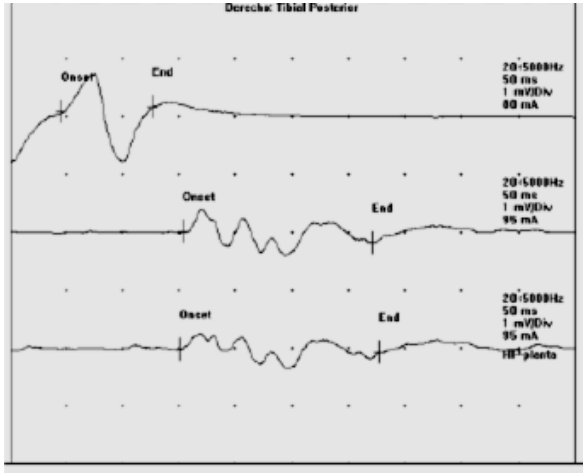


Mario PCID abril 2011-IgG IV

Velocidad de conducción nerviosa motora						
	Comienzo (ms)	Duración (ms)	Amplitud (mV)	Area (µVs)	Distancia (cm)	Velocidad (m/s)
Derecho: Peroneo Hueco Popliteo-pedio						
1.Maleolo- Pedio	5,6	4,7	1,1	1,8	4,0	
2.Cabeza Peronéo	12,6	19,0	0,6	2,5	17,5	25,0
3.RespuestaF-CP-Pedio	13,9	19,4	0,7	3,1	4,0	30,8

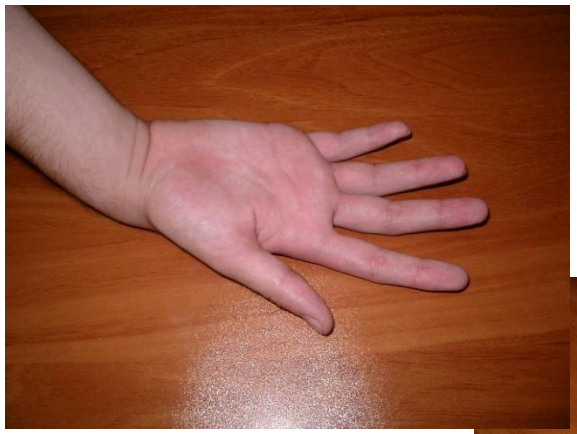


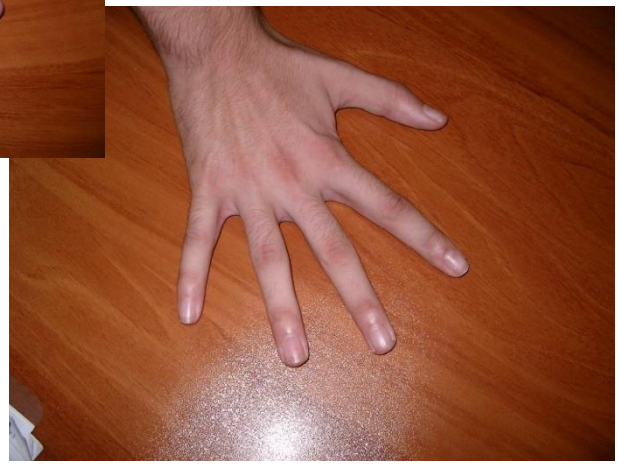
Velocidad de conducción nerviosa motora						
	Comienzo (ms)	Duración (ms)	Amplitud (mV)	Area (µVs)	Distancia (cm)	Velocidad (m/s)
Derecho: Tibial Posterior HP-planta						
1.Maleolo- Planta	4,6	8,1	1,5	3,3	8,0	
2.Hueco Popliteo	15,3	16,8	0,8	3,0	20,0	18,7
3.RespuestaF-HP-Planta	15,0	17,7	0,6	2,5		

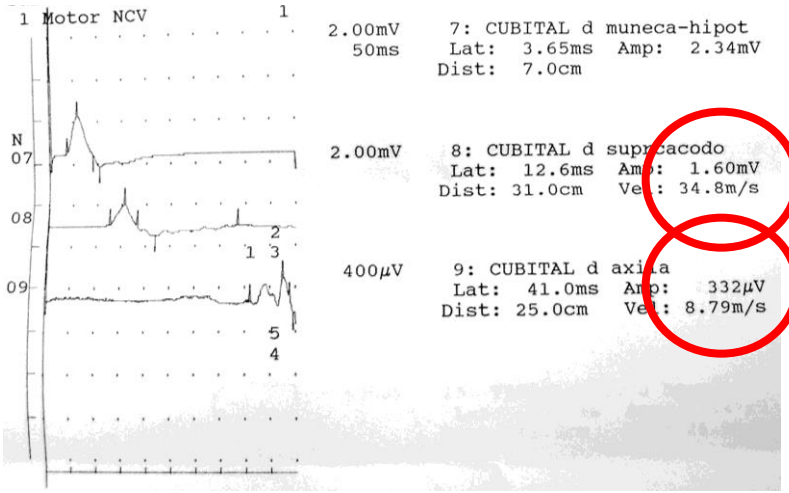


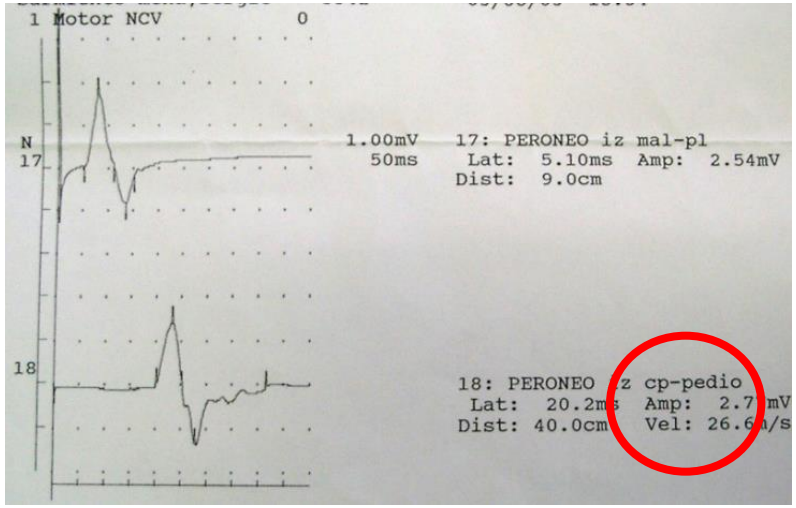
Polineuritis cronica inflam desmielinizante

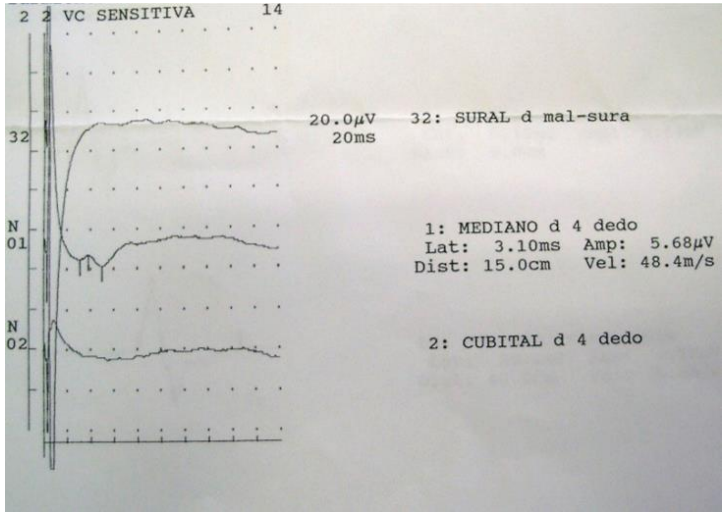
Sergio 558 y 611











POLINEUROPATIA CRONICA INFLAMATORIA DESMIELINIZANTE

RM MEDULAR

Engrosamiento de raices nerviosas

En 38% de los casos

Neuromuscul Disord. 2013;23:103-11.

POLINEUROPATIA CRONICA INFLAMATORIA DESMIELINIZANTE

TRATAMIENTOS

IgG iv
Corticoides

Inmunosupresores
- Azatioprina,
  - De valor discutido en adultos
  - En niños es el más usado, combinado con corticoides.
- Methotrexate
- Ciclofosfamida
- Ciclosporina : sin efecto en dos series.

Micofenolato mofetil

Neuromuscul Disord. 2013;23:103-11.

Intravenous immunoglobulin versus intravenous methylprednisolone for chronic inflammatory demyelinating polyradiculoneuropathy: a randomised controlled trial

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Summary

Background Intravenous immunoglobulin (IVIg) and corticosteroids are effective as initial treatment in patients with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), but little is known about the comparative risk–benefit profile of their long-term use in this disease. We compared the efficacy and tolerability of 6-month therapy with IVIg versus that with intravenous methylprednisolone.

Methods We did a multicentre, randomised, double-blind, placebo controlled, parallel-group study in patients with CIDP. We assessed efficacy and tolerability of IVIg (0·5 g/kg per day for 4 consecutive days) and intravenous methylprednisolone (0·5 g in 250 mL sodium chloride solution per day for 4 consecutive days) given every month for 6 months. Eligible patients had to be in an active or stationary phase of the disease. Allocation to treatment was centrally managed with a computer-generated, 1:1 randomisation scheme with a sequential block size of four. All patients and assessors were unaware of the treatment assignment. After therapy discontinuation, patients were followed up for 6 months to assess relapses. The primary outcome was the difference in the number of patients discontinuing either therapy owing to inefficacy or intolerance. Secondary endpoints included the difference in the proportion of patients experiencing adverse events or worsening after therapy discontinuation. This study is registered with EUDRACT, number 2005-001136-76.

Findings 45 patients (24 IVIg, 21 intravenous methylprednisolone) completed the study; one was excluded for inappropriate inclusion. More patients stopped methylprednisolone (11 [52%] of 21) than IVIg (three [13%] of 24; relative risk 0·54, 95% CI 0·34–0·87; p=0·0085). When adjusted for sex, age, disease duration, comorbidity, modified Rankin scale and ONLS scores at enrolment, and previous treatment with IVIg and steroids, the difference between the two groups remained significant (odds ratio 7·7, 95% CI 1·7–33·9; p=0·0070). Reasons for discontinuation were lack of efficacy (eight in the methylprednisolone group *vs* three in the IVIg group), adverse events (one in the methylprednisolone group), or voluntary withdrawal (two in the methylprednisolone group). Two patients on IVIg died during follow-up after the 6-month assessment. The proportion of patients with adverse events did not differ between the intravenous methylprednisolone group (14 [67%] of 21) and the IVIg group (11 [46%] of 24; p=0·1606). After therapy discontinuation, more patients on IVIg worsened and required further therapy (eight [38%] of 21) than did those on methylprednisolone (none of ten; p=0·0317).

Interpretation Treatment of CIDP with IVIg for 6 months was less frequently discontinued because of inefficacy, adverse events, or intolerance than was treatment with intravenous methylprednisolone. The longer-term effects of these treatments on the course of CIDP need to be addressed in future studies.

Lancet Neurol 2012; 11: 493–502

Neuromuscul Disord. 2013 Feb;23(2):103-11.

Childhood chronic inflammatory demyelinating polyradiculoneuropathy: Combined analysis of a large cohort and eleven published series

Hugh J. McMillan [a], Peter B. Kang [b], H. Royden Jones [b,c], Basil T. Darras [b,*]

Abstract

The clinical presentation, disease course, response to treatment, and long-term outcome of thirty childhood chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) patients are presented representing the largest cohort reported to date. Most children (60%) presented with chronic (>8-weeks) symptom-onset while a smaller proportion showed sub-acute (4–8 weeks) or acute ("GBS-like"; <4 weeks) onset of disease. No gender predilection was observed. The majority of patients had a relapsing (70%) versus a monophasic (30%) temporal profile. Most received initial IVIG monotherapy; 80% showing a good response. Long-term follow-up (mean = 3.8 years) was available for 23 patients; 45% were off all immunomodulatory medications, demonstrating no detectable (55%) or minimal (43%) clinical deficits. Our data were compared with 11 previously published childhood CIDP series providing a comprehensive review of 143 childhood CIDP cases. The combined initial or first-line treatment response across all studies was favourable for IVIG (79% patients) and corticosteroids (84% patients). Response to first-line plasma exchange was poor (only 14% patients improved) although it may offer some transient or partial benefit as an adjuvant or temporary therapy for selected patients. The combined long-term outcome of our cohort and the literature reveals a favourable prognosis for most patients. The combined modified Rankin scale decreased from 3.7 (at presentation) to 0.7 (at last follow-up). This review provides important data pertaining to clinical course, treatment response and long-term outcome of this relatively uncommon paediatric autoimmune disease.

ESCALA MODIFICADA DE RANKIN (MRS)

escala fiable y válida para cuantificar el déficit clínico:

0 = asintomático
1 = sintomas leves que no interfieren con trabajo, colegio o cualquier actividad de vida diaria.
2 = ligera discapacidad (no puede hacer una o más actividades) pero puede ejercer sus AVD personales (vestido, aseo,comida, escritura..).
3 = síntomas moderados (puede andar sin ayudas o con un bastón o muleta), pero requiere ayuda para tareas propias de su edad (comida, aseo, vestido..)
4 = sintomas moderados o graves (no puede andar, silla de ruedas), ni es autonomo para sus AVD.
5 = síntomas, discapacidad graves (en cama, o requiere cuidado constante de otra persona), puede necesitar ventilación mecánica.
6 = muerte.

Rossignol et al:Pediatr Neurol 2007;36(2):88–94.
Simmons et al. Muscle Nerve 1997;20(12):1569–75.

Childhood chronic inflammatory demyelinating polyradiculoneuropathy: Combined analysis of a large cohort and eleven published series

Hugh J. McMillan [a], Peter B. Kang [b], H. Royden Jones [b,c], Basil T. Darras [b,*]

Table 1
Childhood CIDP: clinical presentation and outcome.

Reference	# Patients (male:female)	Mean age onset (years)	Disease course	Modified Rankin score	
				Initial	Follow-up
Current series	30 (13M:17F)	7.6 (1.5–19)	21 Relapsing: 9 monophasic	2.8	0.5[a]
Rossignol et al. [9]	13 (9M:4F)	9 (3–14)	10 Relapsing: 3 monophasic	3.0	1.3
Ryan et al. [10]	16 (5M:11F)	6.3 (2.2–13.8)	6 Relapsing: 10 monophasic[b]	3.4	0.25
Hattori et al. [11]	10 (6M:4F)	11 (2–16)	7 Relapsing : 3 monophasic	4.4	1.9
Simmons et al. [12]	15 (7M:8F)	11.5 (3–17)	10 Relapsing : 2 monophasic[c]	3.5	0.2[c]
Korinthenberg [14]	21 (12M:9F)	8.6 (2–14)	9 Relapsing : 12 monophasic	NR[d]	NR[d]
Nevo et al. [2]	13 (8M:5F)	6.5 (1–16)	10 Relapsing : 3 monophasic	NR	NR
Vedanarayanan et al. [15]	4 (1M:3F)	7.5 (7–9)	3 Relapsing : 1 monophasic	NR	NR
Rodriguez-Casero et al. [16]	5 (3M:2F)	8.0 (4.5–13.9)	0 Relapsing : 5 monophasic	NR	NR
Uncini et al. [17]	5 (1M:4F)	7 (6–11)	NR	NR	NR
Sladky et al. [1]	6 (5M:1F)	NR	NR	NR	NR
Colan et al. [18]	5 (3M:2F)	9.6 (5–17)	3 Relapsing ; 2 monophasic	NR	NR
Total	143 (73M:70F)		79 (61%) Relapsing ; 50 (39%) monophasic		

M, male; F, female; NR, not reported.
[a] Long term follow-up available for 20/30 patients.
[b] Monophasic and progressive were grouped together.
[c] Long term follow-up and treatment data available for 12/15 patients.
[d] Different functional scoring system was used.

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Childhood chronic inflammatory demyelinating polyradiculoneuropathy: Combined analysis of a large cohort and eleven published series

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Table 2
Childhood CIDP: treatment success with initial or first-line therapy.

Reference	# Patients	Patients showing GOOD response (%)		
		IVIG	PE	Corticosteroids
Current series	29[a]	20/25[a] (80%)	0/2 (0%)	Not used[a]
Rossignol et al. [9]	13	2/3 (67%)	Not used	8/10 (80%)
Ryan et al. [10]	16	3/4 (75%)	0/1 (0%)	7/11 (64%)
Hattori et al. [11]	10	1/2 (50%)	1/2 (50%)	4/6 (66%)
Simmons et al. [13]	12	5/6 (83%)	0/2 (0%)	4/4 (100%)
Korinthenberg [14]	21	10/12 (83%)	Not used	8/11 (73%)
Nevo et al. [2]	13	Not used	Not used	13/13 (100%)
Rodriguez-Casero et al. [16]	5[b]	Not used	Not used	3/3 (100%)[b]
Uncini et al. [17]	5	Not used	Not used	4/5 (80%)
Sladky et al. [1]	6	Not used	Not used	6/6 (100%)
Colan et al. [18]	5	Not used	Not used	5/5 (100%)
Total	135	41/52 (79%)	1/7 (14%)	62/74(84%)

Note: Korinthenberg et al. [14]; 1 patient appears to have received combined initial IVIG and corticosteroids although this cannot be confirmed in the text.

[a] Of 30 patients: 1 patient received no treatment at her initial presentation; 2 patients received combined initial IVIG and corticosteroids (therefore, not included in this table).

[b] Of 5 patients: 2 patients received combined initial IVIG and corticosteroids (therefore, not included in this table).

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Childhood chronic inflammatory demyelinating polyradiculoneuropathy: Combined analysis of a large cohort and eleven published series

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Table 3
Childhood CIDP: treatment success to therapies used at any stage of disease.

Reference	# Patients	Patients showing GOOD response (%)		
		IVIG	PE	Corticosteroids
Current series	30	23/29 (68%)	2/5 (16%)	10/10 (100%)
Rossignol et al. [9]	13	3/5 (60%)	1/2 (50%)	8/11 (73%)
Ryan et al. [10]	16	4/6 (67%)	1/4 (25%)	8/12 (67%)
Hattori et al. [11]	10	2/3 (66%)	2/3 (66%)	6/8 (75%)
Simmons et al. [13]	12	7/8 (88%)	2/4 (50%)	5/7 (71%)
Korinthenberg [14]	21	10/12 (83%)	3/5 (60%)	12/20 (60%)
Nevo et al. [2]	13	3/6 (50%)	1/3 (33%)	13/13 (100%)
Vedanarayanan et al. [15]	4	4/4 (100%)	1/3 (33%)	3/4 (75%)
Rodriguez-Casero et al. [16]	5			5/5 (100%)
Uncini et al. [17]	5			4/5 (80%)
Sladky et al. [1]	6			6/6 (100%)
Colan et al. [18]	5			5/5 (100%)
Total	140	56/73 (77%)	13/29 (45%)	85/105 (80%)

Note: These data include response to first-line (initial), second or third line treatments. Treatment can include monotherapy or combined therapies.

Neuromuscul Disord. 2013 Feb;23(2):103-11.

Polineuritis crónica desmielinizante en niños. Pronóstico

- Algo mejor que en adultos

- Dos grupos:

  Progresión corta, 1-3 meses →

  remisión completa en 75%, secuelas leves en 25%

  Progresión larga, prolongada >3 meses :

  Secuelas leves 78%

  Secuelas graves 22%

Rossignol et al. Pediatr Neurol 2007;36:88–94.