

# Neurology Condition Assessment Methods

## Guillain-Barre´ syndrome (GBS) Disability Score

The Guillain-Barré syndrome (GBS) disability score is a widely accepted scoring system to assess the functional status of patients with GBS. It was originally described in Hughes et al. (1978) and since then, various iterations have appeared in the literature. The adaptation best suited for use in the *Criteria* and BloodSTAR is from van Koningsveld et al (2007). The *Criteria* requires that the patient's level of disability be documented using the scale from 0 to 6 as below.

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

Reproduced from Lancet Neurol 2007, Vol.6, [van Koningsveld R, Steyerberg EW, Hughes RAC, Swan AV, van Doorn PA, Jacobs BC.](#) A clinical prognostic scoring system for Guillain-Barré syndrome, pp. 589-94, Copyright (2007), with permission from Elsevier.

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## Modified Rankin Scale (MRS)

The Modified Rankin Scale was originally developed by Rankin J (1957) and has been replicated for use in CIDP, AMAE, Stiff person syndrome and Susac syndrome in the *Criteria* from Van Swieten et al. (1987). It also appears on the [Stroke Society of Australia](#) website. The *Criteria* requires the patient's level of disability to be assessed using a scale from 0 to 6 ranging from no symptoms at all to death.

The Modified Rankin Scale from which this scale has been replicated is available on the [Stroke Society of Australia](#) website at:

- [http://www.strokesociety.com.au/index.php?option=com\\_content&view=article&id=292:modified-rankin-scale-astn&catid=40:astn](http://www.strokesociety.com.au/index.php?option=com_content&view=article&id=292:modified-rankin-scale-astn&catid=40:astn)

Please note for the conditions paediatric autoimmune neuropsychiatric disorder associated with streptococcal infections (PANDAS) or paediatric acute neuropsychiatric disorders (PANS), Sjogren's syndrome and Rasmussen syndrome the description of disabilities in the Modified Rankin Scale has been adapted given that there is no loss of motor function.

For further information please see the following articles:

- Bonita R, Beaglehole R. Recovery of motor function after stroke. *Stroke* 1988 19(12):1497-1500
- Stroke Society of Australia, The Modified Rankin Scale  
[http://www.strokesociety.com.au/index.php?option=com\\_content&view=article&id=292:modified-rankin-scale-astn&catid=40:astn](http://www.strokesociety.com.au/index.php?option=com_content&view=article&id=292:modified-rankin-scale-astn&catid=40:astn)
- Rankin J. Cerebral vascular accidents in patients over the age of 60. *Scott Med J* 1957; 2: 200-15
- Van Swieten JC, Koudstaal PJ, Visser MC, Schouten HJ, van Gijn J. Interobserver agreement for the assessment of handicap in stroke patients. *Stroke* 1987. 19(5).604-607

## Medical Research Council (MRC) Scale for Muscle Strength

The Medical Research Council (MRC) Scale for Muscle Strength is a commonly used scale for assessing muscle strength from Grade 5 (normal) to Grade 0 (no visible contraction). It was originally described by the [Medical Research Council in 1943](#). The MRC sum score was first described by Kleyweg et al (1988) for use in the Dutch Guillain-Barré trial. This score was defined as the sum of MRC scores from six muscles in the upper and lower limbs on both sides so that the score ranged from 60 (normal) to 0 (quadriplegic).

The *Criteria* requires that each of the six muscle groups listed in the table are examined bilaterally, each with a score from 0 to 5 according to the scale in the right hand column.

MRC Sum score			
Muscle		Score 0-5	MRC scale for muscle strength (0-5)
Shoulder abductors	Left		Grade 5: Normal  Grade 4: Movement against gravity and resistance  Grade 3: Movement against gravity over (almost) the full range  Grade 2: Movement of the limb but not against gravity  Grade 1: Visible contraction without movement of the limb (not existent for hip flexion)  Grade 0: No visible contraction  MRC grade for each muscle given in full numbers: (4+/4.5 =4) (4- =3) (5- = 4)
	Right		
Elbow flexors	Left		
	Right		
Wrist extensors	Left		
	Right		
Hip flexors	Left		
	Right		
Knee extensors	Left		
	Right		
Foot dorsiflexors	Left		
	Right		
<b>Total (out of 60)</b>			

Reproduced from Muscle Nerve 1991, Vol.14 (11), [Kleyweg RP, van der Meche FGA, Schmitz PIM. Interobserver agreement in the assessment of muscle strength and functional abilities in Guillain-Barré syndrome](#), pp. 1103-1109, Copyright © 2004, John Wiley and Sons.

Special credit - [Medical Research Council. 1976. Aids to the Examination of the Peripheral Nervous System \(Memorandum No. 45\)](#). The MRC Muscle scale is licensed by the Medical Research Council under the Open Government License.

## Childhood Myositis Assessment Scale – CMAS

The Childhood Myositis Assessment Scale (CMAS) has been shown to be a valid measure of physical function in children with juvenile idiopathic inflammatory myopathies. It is the most commonly used assessment tool and has been recommended as a core set measure. The CMAS was first published in 1999 by Lovell et al, however the standardized method for conducting the scale can be found below and at the International Myositis Assessment and Clinical Studies Group web site.

The *Criteria* requires submission of a score calculated by assessing each of the 14 manoeuvres in the table and scored according the scale to yield a total maximum score of 52.

Childhood Myositis Assessment Scale scoring sheet	
<p><b>1. Head elevation (neck flexion):</b>            0 = Unable            1 = 1–9 seconds            2 = 10–29 seconds            3 = 30–59 seconds            4 = 60–119 seconds            5 = ≥2 minutes</p> <p style="text-align: right;">No. of seconds _____</p>	Score
<p><b>2. Leg raise/touch object:</b>            0 = Unable to lift leg off table            1 = Able to clear table, but cannot touch object            2 = Able to lift leg high enough to touch object</p>	Score
<p><b>3. Straight leg lift/duration:</b>            0 = Unable            1 = 1–9 seconds            2 = 10–29 seconds            3 = 30–59 seconds            4 = 60–119 seconds            5 = ≥2 minutes</p> <p style="text-align: right;">No. of seconds _____</p>	Score
<p><b>4. Supine to prone:</b>            0 = Unable. Has difficulty even turning onto side; able to pull arms under torso only slightly or not at all            1 = Turns onto side fairly easily, but cannot fully free arms and is not able to fully assume a prone position            2 = Easily turns onto side; has some difficulty freeing arms, but fully frees them and fully assumes a prone position            3 = Easily turns over, fully frees right arm with no difficulty</p>	Score
<p><b>5. Sit-ups:</b>            For each type of sit-up enter either “0” (unable) or “1” (able). Then enter the total sit-up score (0-6).</p> <p>Hands on thighs, with counterbalance _____            Hands across chest, with counterbalance _____            Hands behind head, with counterbalance _____            Hands on thighs, without counterbalance _____            Hands across chest, without counterbalance _____            Hands behind head, without counterbalance _____</p>	Score
<p><b>6. Supine to sit:</b>            0 = Unable by self            1 = Much difficulty. Very slow, struggles greatly, barely makes it. Almost unable            2 = Some difficulty. Able, but is somewhat slow, struggles some            3 = No difficulty</p>	Score

<p><b>7. Arm raise/straighten:</b>  0 = Cannot raise wrists  1 = Can raise wrists at least up to the level of the acromioclavicular joint, but not above top of head  2 = Can raise wrists above top of head, but cannot raise arms straight above head so that elbows are in full extension  3 = Can raise arms straight above head so that elbows are in full extension</p>	Score
<p><b>8. Arm raise/duration:</b>  Can maintain wrists above top of head for:  0 = Unable  1 = 1–9 seconds  2 = 10–29 seconds  3 = 30–59 seconds  4 = ≥ 60 seconds</p> <p style="text-align: right;">No. of seconds _____</p>	Score
<p><b>9. Floor sit:</b>  Going from a standing position to a sitting position on the floor  0 = Unable. Afraid to even try, even if allowed to use a chair for support. Child fears that he/she will collapse, fall into a sit, or harm self  1 = Much difficulty. Able, but needs to hold onto a chair for support during descent. (Unable or unwilling to try if not able to use a chair for support)  2 = Some difficulty. Can go from stand to sit without using a chair for support, but has at least some difficulty during descent. Descends somewhat slowly and/or apprehensively; may not have full control or balance as manoeuvres into a sit  3 = No difficulty. Requires no compensatory manoeuvring</p>	Score
<p><b>10. All-fours manoeuvre:</b>  0 = Unable to go from a prone to an all-fours position  1 = Barely able to assume and maintain an all-fours position  2 = Can maintain all-fours position with straight back and head raised (so as to look straight ahead). But, cannot creep (crawl) forward  3 = Can maintain all-fours, look straight ahead, and creep (crawl) forward  4 = Maintains balance while lifting and extending leg</p>	Score
<p><b>11. Floor rise:</b>  Going from a kneeling position on the floor to a standing position  0 = Unable, even if allowed to use a chair for support  1 = Much difficulty. Able, but needs to use a chair for support. Unable if not allowed to use a chair  2 = Moderate difficulty. Able to get up without using a chair for support, but needs to place one or both hands on thighs/knees or floor. Unable without using hands  3 = Mild difficulty. Does not need to place hands on knees, thighs, or floor, but has at least some difficulty during ascent  4 = No difficulty</p>	Score
<p><b>12. Chair rise:</b>  0 = Unable to rise from chair, even if allowed to place hands on sides of chair seat  1 = Much difficulty. Able, but needs to place hands on sides of seat. Unable if not allowed to place hands on knees/thighs  2 = Moderate difficulty. Able, but needs to place hands on knees/thighs. Does not need to place hands on side of seat  3 = Mild difficulty. Able; does not need to use hands at all, but has at least some difficulty  4 = No difficulty</p>	Score
<p><b>13. Stool step:</b>  0 = Unable</p>	Score

<p>1 = Much difficulty. Able, but needs to place one hand on exam table or examiner's hand</p> <p>2 = Some difficulty. Able; does not need to use exam table for support, but needs to use hand(s) on knee/thigh</p> <p>3 = Able. Does not need to use exam table or hand(s) on knee/thigh</p>	
<p><b>14. Pick up:</b></p> <p>0 = Unable to bend over and pick up pencil off floor</p> <p>1 = Much difficulty. Able, but relies heavily on support gained by placing hand(s) on knees/thighs</p> <p>2 = Some difficulty. Needs to at least minimally and briefly place hand(s) on knees/thighs for support and is somewhat slow</p> <p>3 = No difficulty. No compensatory manoeuvre necessary</p>	Score
<b>Total score (maximum possible score 52)</b>	

Reproduced from *Arthritis Care Res* 2014, Vol.66, [Huber, A, Lovell, D.J, Pilkington, C.A et al., Confusion Concerning Multiple Versions of the Childhood Myositis Assessment Scale](#), p.648, Copyright (2007), with permission from John Wiley and Sons. Copyright © 2014 by the American College of Rheumatology

## The Myasthenia Gravis Composite score – MGC score

The Myasthenia Gravis Composite score has been validated as an outcome measure of signs and symptoms for patients with myasthenia gravis (MG). The score was first described by Burns et al. (2008) and has been replicated for the use in MGC in the *Criteria* from Burns et al. (2010). The *Criteria* requires submission of a score calculated by completing each section in the table after assessment of both physician examination and patient reported signs and symptoms.

The Myasthenia Gravis Composite score	Score
<b>Ptosis, upward gaze (physician examination)</b> 0 = >45 seconds 1 = 11-45 seconds 2 = 1-10 seconds 3 = immediate	Score
<b>Double vision on lateral gaze, left or right (physician examination)</b> 0 = >45 seconds 1 = 11-45 seconds 3 = 1-10 seconds 4 = immediate	Score
<b>Eye closure (physician examination)</b> 0 = Normal 0 = Mild weakness (can be forced open with effort) 1 = Moderate weakness (can be forced open easily) 2 = Severe weakness (unable to keep eyes closed)	Score
<b>Talking (patient history)</b> 0 = Normal 2 = Intermittent slurring or nasal speech 4 = Constant slurring or nasal but can be understood 6 = Difficult to understand speech	Score
<b>Chewing (patient history)</b> 0 = Normal 2 = Fatigue with solid food 4 = Fatigue with soft food 6 = Gastric tube	Score
<b>Swallowing (patient history)</b> 0 = Normal 2 = Rare episode of choking or trouble swallowing 5 = Frequent trouble swallowing, eg. necessitating changes in diet 6 = Gastric tube	Score
<b>Breathing (thought to be caused by MG)</b> 0 = Normal 2 = Shortness of breath with exertion 4 = Shortness of breath at rest 9 = Ventilator dependence	Score
<b>Neck flexion or extension (weakest, physician examination)</b> 0 = Normal 1 = Mild weakness 3* = Moderate weakness (ie. ~50% weak, ±15%) 4 = Severe weakness	Score
<b>Shoulder abduction (physician examination)</b> 0 = Normal 2 = Mild weakness	Score

4* = Moderate weakness (ie.~50% weak, ±15%) 5 = Severe weakness	
<b>Hip flexion (physician examination)</b> 0 = Normal 2 = Mild weakness 4* = Moderate weakness (ie.~50% weak, ±15%) 5 = Severe weakness	Score
<b>Total score (maximum possible score 50)</b>	

\*Moderate weakness for neck and limb items should be construed as weakness that equals roughly 50% plus or minus 15% of expected normal strength. Any weakness milder than that would be mild and any weakness more severe than that would be classified as severe.

The Myasthenia Gravis Composite score replicated from [Burns TM, Conaway M, Sanders DB. The MG composite: a valid and reliable outcome measure for myasthenia gravis. Neurology 2010 May 4; 74\(18\): 1434–40](#) with permission from T.M.Burns.



## Expanded Disability Status Scale (EDSS)

The Expanded Disability Status Scale (EDSS) is a commonly used scale for assessing the level of disability in people with multiple sclerosis. The scale was originally described by Kurtze (1983) and has been adapted on the [Multiple Sclerosis Trust](#) website.

The *Criteria* requires that each of the functional systems listed below be reviewed. The description in the table below should be referred to and for ambulant patients a score from 0 (no disability) to 5.5 (more severe disability) be indicated. If the patient has impairment such that assistance is required to enable walking (cane, crutches, frame) the score will be from 0 to 6.5, if a wheelchair is required for all but a few steps the score will be 7.0 – 8.5, if confined to bed the score will be 9 and death due to MS is scored 10.

### Functional systems

- pyramidal - weakness or difficulty moving limbs
- cerebellar - ataxia, loss of coordination or tremor
- brainstem - problems with speech, swallowing and nystagmus
- sensory - numbness or loss of sensations
- bowel and bladder function
- visual function
- cerebral (or mental) functions
- other

ALL functional system scores must be scored individually and then the EDSS calculated from the table below.

<b>Expanded Disability Status Scale (EDSS)</b>	
<b>Score</b>	<b>Description</b>
<b>0.0</b>	No disability
<b>1.0</b>	No disability, minimal signs in one FS
<b>1.5</b>	No disability, minimal signs in more than one FS
<b>2.0</b>	Minimal disability in one FS
<b>2.5</b>	Mild disability in one FS or minimal disability in two FS
<b>3.0</b>	Moderate disability in one FS, or mild disability in three or four FS. No impairment to walking
<b>3.5</b>	Moderate disability in one FS and more than minimal disability in several others. No impairment to walking
<b>4.0</b>	Significant disability but self-sufficient and up and about some 12 hours a day. Able to walk without aid or rest for 500m
<b>4.5</b>	Significant disability but up and about much of the day, able to work a full day, may otherwise have some limitation of full activity or require minimal assistance. Able to walk without aid or rest for 300m
<b>5.0</b>	Disability severe enough to impair full daily activities and ability to work a full day without special provisions. Able to walk without aid or rest for 200m
<b>5.5</b>	Disability severe enough to preclude full daily activities. Able to walk without aid or rest for 100m
<b>6.0</b>	Requires a walking aid - cane, crutch, etc. - to walk about 100m with or without resting
<b>6.5</b>	Requires two walking aids - pair of canes, crutches, etc. - to walk about 20m without resting
<b>7.0</b>	Unable to walk beyond approximately 5m even with aid. Essentially restricted to wheelchair; though wheels self in standard wheelchair and transfers alone. Up and about in wheelchair some 12 hours a day
<b>7.5</b>	Unable to take more than a few steps. Restricted to wheelchair and may need aid in transferring. Can wheel self but cannot carry on in standard wheelchair for a full day and may require a motorised wheelchair
<b>8.0</b>	Essentially restricted to bed or chair or pushed in wheelchair. May be out of bed itself much of the day. Retains many self-care functions. Generally has effective use of arms
<b>8.5</b>	Essentially restricted to bed much of day. Has some effective use of arms retains some self-care functions

<b>9.0</b>	Confined to bed. Can still communicate and eat
<b>9.5</b>	Confined to bed and totally dependent. Unable to communicate effectively or eat/swallow
<b>10.0</b>	Death due to MS

The Expanded Disability Status Scale has been replicated from the Multiple Sclerosis Trust website at:

- <https://www.mstrust.org.uk/a-z/expanded-disability-status-scale-edss> with permission from the Multiple Sclerosis Trust UK

## Cerebellar Functional System Score

The Cerebellar Functional System Score has been adapted from the larger Functional Systems Score first described by Kurtzke (1983) which was designed for assessing people with multiple sclerosis. However, the Cerebellar Functions section can be used to demonstrate initial disability and response to Ig therapy in children with Opsoclonus-myoclonus ataxia (OMA).

The *Criteria* requires that the patient's impairment is indicated using the scale from 0 to 5 as below.

Cerebellar Functional System Score	
Score	Criteria
0	Normal
1	Abnormal signs without disability
2	Mild ataxia
3	Moderate truncal or limb ataxia
4	Severe ataxia, all limbs
5	Unable to perform coordinated movements due to ataxia
X	Used throughout after each number when weakness (grade 3 or more on pyramidal) interferes with testing

The Cerebellar Functional System Score is a subset of the Expanded Disability Status Scale which has been replicated from the Multiple Sclerosis Trust website at: <https://www.mstrust.org.uk/a-z/expanded-disability-status-scale-edss> with permission from the Multiple Sclerosis Trust UK

## Overall Neuropathy Limitations Scale

The Overall Disability Sum Score (ODSS) was the first scale designed to assess the limitations of patients with immune-mediated peripheral neuropathies. The ODSS focuses on upper and lower limb functions, and consists of a checklist for interviewing patients. The ODSS has showed reliability, responsiveness and construct validity in people with Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy and paraprotein-associated demyelinating neuropathy. To reduce a possible ceiling effect, the ODSS was modified slightly to include climbing stairs and running. This new measure is called the Overall Neuropathy Limitations Scale (ONLS). The *Criteria* requires that this scale be completed by adding the total of the Arm scale score (0-5) and Leg scale score (0-7) yielding a total score of 0-12.

To evaluate a change in disability on patient review an Adjusted ONLS is recorded. The adjusted score is identical to the ONLS disability score except for the exclusion of changes in upper limb function from 0 to 1 or from 1 to 0, because these changes have not been judged to be clinically significant in all patients. All other 1-point steps in either the arm or leg scale represent clinically meaningful changes in disability.

Overall Neuropathy Limitations Scale			
<b>Instructions:</b> The examiner should question <b>and</b> observe the patient in order to determine the answers to the following questions. Note should be made of any other disorder other than peripheral neuropathy which limits function.			
<b>Arms Scale</b>			
Does the patient have any symptoms in their hands or arms, e.g. tingling, numbness or weakness? (if no, go to legs section)	Yes		No
Is the patient affected in their ability to:	Not affected	Affected but not prevented	Prevented
Wash and brush their hair	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Turn a key in a lock	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Use a knife and fork together (or spoon, if knife and fork not used)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Do or undo buttons or zips	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Dress the upper part of their body excluding buttons or zips	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If all these functions are prevented can the patient make purposeful movements with their hands or arms?	Yes	No	Not applicable
<b>Arms grade score</b>			
0= Normal			
1= Minor symptoms in one or both arms but not affecting any of the functions listed			
2= Disability in one or both arms affecting but not preventing any of the functions listed			
3= Disability in one or both arms preventing at least one but not all functions listed			
4= Disability in both arms preventing all functions listed but purposeful movement still possible			
5= Disability in both arms preventing all purposeful movements			
<b>Legs Scale</b>			
	Yes	No	Not applicable
Does the patient have difficulty running or climbing stairs?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Does the patient have difficulty with walking?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Does their gait look abnormal?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
How do they mobilise for about 10 metres (i.e. 33 feet)?			
Without aid	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
With one stick or crutch or holding to someone's arm	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
With two sticks or crutches or one stick or crutch holding onto someone's arm or frame	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
With a wheelchair	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If they use a wheelchair, can they stand and walk 1 metre with the help of one person?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
If they cannot walk as above are they able to make some purposeful movements of their legs, e.g. reposition legs in bed?	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Does the patient use ankle foot orthoses/braces? ( If yes, please indicate, Right	<input type="checkbox"/>	<input type="checkbox"/>	

or Left )			
<b>Legs grade score</b>			
0= Walking/climbing stairs/running not affected			
1= Walking/climbing stairs/running is affected, but gait does not look abnormal			
2= Walks independently but gait looks abnormal			
3= Requires unilateral support to walk 10 metres (stick, single crutch, one arm)			
4= Requires bilateral support to walk 10 metres (sticks, crutches, crutch and arm, frame)			
5= Requires wheelchair to travel 10 metres but able to stand and walk 1 metre with the help of one person			
6= Restricted to wheelchair, unable to stand and walk 1 metre with the help of one person, but able to make some purposeful leg movements			
7= Restricted to wheelchair or bed most of the day, unable to make any purposeful movements of the legs			
Is there any disorder, other than peripheral neuropathy, which affects the above functions? If Yes, please describe in the comments section in BloodSTAR.		Yes	No
<b>Overall Neuropathy Limitation Scale Score</b>			
<b>Arm scale score (0 to 5)</b>	/5	<b>+ Leg scale score (0 to 7)</b>	/7
Range: 0 (no disability) to 12 (maximum disability).			
<b>Total ONLS score (please enter into BloodSTAR): /12</b>			

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## The Six Minute Walk Test – 6MWT

The Six Minute Walk Test (6MWT) is an objective evaluation of functional capacity, which has been demonstrated to be accurate, reproducible, simple to administer and well-tolerated. Relevance to neuromuscular diseases has been established. Guidelines exist to standardise the performance of the 6MWT with adaptations and standard reference values described for use in children.

**Instructions:** The *Criteria* requires documentation of the distance walked in 6 minutes using a standardised methodology. For methodological instructions please see the American Thoracic Society

**ATS Statement: Guidelines for the Six-Minute Walk Test**

<https://www.thoracic.org/statements/resources/pfet/sixminute.pdf>

Detailed instructions are available in:

ATS Statement (2002): Guidelines for the Six-Minute Walk Test. *Am J Respir Crit Care Med* 166:111

## Distribution of Stiffness index

The Distribution of Stiffness Index was originally described in [Dalakas et al. \(2000\)](#) and replicated for the use in Stiff Person Syndrome in the *Criteria* from Dalakas et al. (2017). It can be used to provide a baseline and to monitor the response to Ig therapy.

The Distribution of Stiffness Index in BloodSTAR has been replicated from:

- Dalakas MC, Rakocevic, G., Dambrosia J.M., Alexopoulos, H. and McElroy, B. A Double-Blind, Placebo-Controlled Study of Rituximab in Patients with Stiff Person Syndrome. *Ann Neurol* 2017;82:271-277 <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5755707/>