

## Supplementary Online Content

Clardy SL, Lennon VA, Dalmau J, et al. Childhood onset of stiff-man syndrome. *JAMA Neurol*. Published online October 7, 2013. doi:10.1001/jamaneurol.2013.4442.

**eTable.** 12 Stiff-Man Syndrome Spectrum Pediatric Patients Previously Reported in the Literature

This supplementary material has been provided by the authors to give readers additional information about their work.

**eTable. 12** Stiff-Man Syndrome Spectrum Pediatric Patients Previously Reported in the Literature

	<b>Bowler 1960<sup>15</sup></b>	<b>Kugelma ss 1961<sup>16</sup></b>	<b>Isaacs 1979<sup>17</sup></b>	<b>Daras and Spiro 1981<sup>18</sup></b>	<b>Maccario et al. 1984<sup>19</sup></b>	<b>Udani et al. 1997<sup>20</sup></b>	<b>Garzo et al. 1998<sup>21</sup></b>	<b>Markandey ulu et al, 2001<sup>22</sup></b>	<b>Mikaeloff et al. 2001<sup>23</sup></b>	<b>Sanefuji et al. 2012<sup>24</sup></b>	<b>Fekete and Jankovic 2012<sup>25</sup></b>	<b>Damasio et al<sup>26</sup></b>
<b>Age at exam (years)</b>	7	11	8	16	16	1	6	11	12	7	12	14
<b>Age at onset (years)</b>	6	10	6	15	13	3 months	6	6	12	7	5	14
<b>Sex</b>	M	M	F	M	F	F	M	M	M	F	M	F
<b>Race/ethnicity</b>	Chinese	N/A	Black	White	N/A	South Asian	N/A	N/A	N/A	N/A	Hispanic	White
<b>Muscle involvement:</b>												
<b>Upper Limbs</b>	+	+	+	+	+	+	-	+	+	-	+	+
<b>Lower limbs</b>	+	+	+	+	+	+	+	+	+	+	+ <sup>1</sup>	+
<b>Trunk</b>	+	+	+	+	+	+	+	-	+	+	+	+
<b>Lumbar hyperlordosis</b>	-	+	+	+	-	+	+	-	+	-	+	+
<b>Head and/or neck</b>	+ <sup>2</sup>	+	-	-	+	+ <sup>2</sup>	-	-	+	-	-	+
<b>Whole body spasms</b>	+	+	+	+	+	+	+	-	+	-	-	+
<b>Respiratory</b>	-	-	Unavailable	Unavailable	+ likely died of respiratory spasm after withdrawal of Baclofen	+	-	-	-	-	-	Tachypnea
<b>DTRs</b>	Normal	Brisk	Reduced	Brisk	Reduced	Normal	Normal	Brisk	Unavailable	Normal	Sustained right ankle clonus	Brisk
<b>Other clinical, radiological findings</b>	Trismus	-	-	Minor right ankle injury precipitated stiffness.	Prominent pupillary dilatation during spasms.	Trismus, blepharospasm.	-	-	MRI brain: T2 signal in right hippocampal region. Normal IQ but new	Antecedent infection, OCD behaviors, lethargy. MRI brain: striatal lesions on	Age 11, nightly awakening with anxiety and tachycardia followed	Onset of symptoms after upper respiratory tract infection; trismus laterocollis

									long-term memory deficit noted at onset.	diffusion imaging and SPECT, resolved in 10 months. EEG: bilaterally synchronous bursts of 2-3 Hz delta, 1sec duration.	by painful lordotic posturing, progressed to several times daily.	myoclonus hyperekplexia urinary retention
<b>Other medical diagnoses</b>	None	None	None	Depression during acute illness.	None	Partial cleft lip	None	None	3 year history of well-controlled DM Type I.	None	Generalized seizure (once, age 5, controlled on levetiracetam)	None
<b>Serological findings</b>	N/A	N/A	N/A	N/A	N/A	N/A	GAD65-IgG not detected in serum and CSF	N/A	GAD65-IgG detected in serum at high titer	GAD65-IgG testing negative in serum	GAD65-IgG detected in serum in high titer (4,405 nmol/l)	GlyRa1-IgG detected in serum and CSF
<b>Electrophysiology</b>	N/A	N/A	Spontaneous motor unit activity at rest, aggravated by touch and movement.	Continuous activity at rest.	Denervation potentials present at rest.	Complex repetitive discharges and continuous motor unit activity	Continuous motor unit activity at rest with superimposed spasms.	Continuous motor unit activity at rest.	Continuous motor unit potentials in paraspinal muscles and limbs.	Continuous motor unit activity at rest.	N/A	Rhythmic bursts of normal muscle action potentials
<b>Good response</b>	No treatment given	ACTH, cortisone	diazepam, baclofen	diazepam	Baclofen, diazepam	Diazepam, baclofen, valproate, steroids	Diazepam, baclofen	Diazepam	IVIg	Diazepam, IVIg, Pulse IV steroids	Rituximab, diazepam, clonazepam, IVIg	IVIg, methylprednisolone, levetiracetam
<b>Poor response</b>	No treatment given	-	glycine, pyridostigmine, phenytoin	-	Valproate, carbamazepine, clonazepam	Carbamazepine, phenytoin	-	-	Baclofen, diazepam, vigabatrin	Baclofen, clonazepam	Baclofen, gabapentin	Clonazepam

					m, steroids							
<b>Duration of follow-up (years)</b>	<1	N/A	N/A	N/A	N/A	4	<1	N/A	<1	3	N/A	2
<b>Course/Outcome</b>	Intermittent/Good		Progressive	Acute/Good	Died	Intermittent/Good	Acute/Good	Intermittent/Good	Subacute/Good	Subacute/Good	Subacute/Good	

**Abbreviations:** ACTH = adrenocorticotrophic hormone; DM= diabetes mellitus; F=female; GAD65=glutamic acid decarboxylase 65kDa isoform; GlyR $\alpha$ 1= glycine receptor  $\alpha$ 1 subunit; IVIg=intravenous immune globulin; M=male; N/A = not available or mentioned; OCD = obsessive compulsive disorders; SPECT = single photon emission computerized tomography.

<sup>1</sup> right lower extremity predominant; <sup>2</sup> risus sardonicus, trismus, blepharospasm and drooling noted