

# Chronic Pain and its Management in Kennedy's Disease

Raghav Govindarajan, MD, FISQua, FACSc, FCPP

Assistant Professor

Department of Neurology

University of Missouri

# Objectives:

1. To understand the clinical spectrum of pain and fatigue in SBMA
2. To identify common causes of pain in SBMA
3. To discuss common treatment strategies

## **Prevalence of pain in SBMA:**

- Pain is an under recognized clinical symptom in neuromuscular disorders
- SBMA is no different, although data on its prevalence are lacking
- In survey that was done on various neuromuscular patients, 40-70% noted pain
- Pain scales varied from 6-8 (on a scale 0-10)
- Pain was present throughout the day in most patients

## Spectrum of Pain:

- Pain is described as “Deep,” “tiring,” “sharp,” and “dull”
- Patients with amyotrophic lateral sclerosis and myotonic muscular dystrophies reported the greatest pain interference
- The most frequent pain site, overall, was back (49%), followed by leg (47%), shoulder (43%), neck (40%), buttock and hip(s) (37%), feet (36%), arm(s) (36%), and hand(s) (35%).
- Most patients had pain at multiple sites (at least 3) some had more 6 sites
- Patients with SBMA typically noted more crampy pain than others

## Pain sites:

**Table 6: Pain Locations Among Subjects With Pain**

Location	% With Pain
Head/face	20
Neck	40
Shoulder(s)	43
Back	49
Chest	17
Arm(s)	36
Hand(s)	35
Buttock/hip(s)	37
Abdomen/pelvis	16
Legs	47
Feet	36

Chest and abdominal pain were least reported by the patients

## Impact of pain on quality of life:

**Table 7: General US and Study Sample (subjects with pain)  
Norms of SF-36 Scores**

SF-36 Scale Score <sup>†</sup>	General US Norms <sup>‡</sup>	Study Sample
Physical functioning	84.15±23.28 <sup>a</sup>	50.41±15.98 <sup>b</sup>
Role-physical	80.96±34.00 <sup>a</sup>	62.28±17.77 <sup>b</sup>
Bodily pain	75.15±20.34 <sup>a</sup>	51.50±15.74 <sup>b</sup>
General health	71.95±20.34 <sup>a</sup>	58.76±20.37 <sup>b</sup>
Vitality	60.86±20.96 <sup>a</sup>	47.95±17.10 <sup>b</sup>
Social functioning	83.28±22.69 <sup>a</sup>	67.12±20.93 <sup>b</sup>
Role-emotional	81.26±33.04	79.01±21.44
Mental health	74.74±18.05	73.23±14.94

Most significant effect of pain was on vitality, followed closely by physical functioning

## Interference with life:

**Table 5: Pain Interference Among Subjects With Pain**

Pain Interference Item*	Mean $\pm$ SD
General activity	3.70 $\pm$ 3.10 <sup>ab</sup>
Mood	3.81 $\pm$ 2.89 <sup>ab</sup>
Mobility (ability to get around)	3.99 $\pm$ 3.34 <sup>b</sup>
Normal work	4.46 $\pm$ 3.39 <sup>c</sup>
Relations with other people	2.56 $\pm$ 2.85 <sup>d</sup>
Sleep	4.04 $\pm$ 3.48 <sup>abc</sup>
Enjoyment of life	3.91 $\pm$ 3.07 <sup>b</sup>
Self-care	2.60 $\pm$ 3.24 <sup>d</sup>
Recreational activities	4.63 $\pm$ 3.59 <sup>c</sup>
Social activities	3.41 $\pm$ 3.21 <sup>a</sup>

Pain had greatest impact on recreational activities, followed by day to day work

## **Causes of pain:**

### Mechanical:

- The pain in the low back and legs may be because of the profound gait abnormalities often associated with muscle weakness
- Many of our patients will have a Trendelenburg gait, with increased lumbar lordosis thus causing significant back pain
- There is also significant shoulder girdle weakness and instability as well, which could explain the upper back and neck and shoulder



## Causes of Pain:

Arthritic pain:

- Because of muscles weakness and tendon/ligament laxity there is increased wear and tear in the joints.
- This predisposes to secondary arthritis and joint deformities
- This further affects gait and walking which in turn predisposes to more pain

## Causes of Pain:

### Neuropathic

- Neuropathic pain includes a spectrum of presentation such as burning, stinging, aching, itching and many more
- Cramps are painful, involuntary muscle contraction
- While there many causes of cramps, in SBMA it is most likely related to the loss of axons
- Similarly significant and severe fasciculation are uncomfortable and rare cases are painful

**Taken together all these different types of pain can cause significant impact on quality of life in patients with SBMA**

## Medical management of pain

Pain treatment	All Subjects With Pain (n=141)		Subjects With Severe Pain (n=38)	
	% Tried/ % Still Use*	Average Relief $\pm$ SD <sup>†</sup>	% Tried/ % Still Use*	Average Relief $\pm$ SD <sup>†</sup>
Ibuprofen, aspirin	61/65	5.22 $\pm$ 2.83	47/78	4.25 $\pm$ 3.05
Acetaminophen	47/58	4.11 $\pm$ 2.93	37/50	3.31 $\pm$ 2.87
Physical therapy	43/42	4.54 $\pm$ 2.66	50/37	3.89 $\pm$ 2.25
Narcotics	35/63	6.37 $\pm$ 2.74	42/56	5.75 $\pm$ 2.79
Massage	34/44	5.48 $\pm$ 2.73	34/39	4.91 $\pm$ 3.36
Neurontin	18/50	4.78 $\pm$ 3.02	24/56	4.57 $\pm$ 3.05
Muscle relaxants	18/60	5.78 $\pm$ 2.88	21/50	4.25 $\pm$ 1.26
Tricyclic antidepressants	15/38	4.53 $\pm$ 3.28	18/29	5.43 $\pm$ 2.99
Acupuncture	11/25	5.29 $\pm$ 3.22	3/100	6.00 $\pm$ 0.00
Magnets	11/25	3.13 $\pm$ 3.16	11/25	1.75 $\pm$ 2.87
Biofeedback/relaxation training	8/55	4.42 $\pm$ 2.50	11/75	4.50 $\pm$ 1.91
Counseling	9/67	4.70 $\pm$ 2.50	18/71	4.17 $\pm$ 2.64
Chiropractic manipulation	4/85	7.33 $\pm$ 3.78	0/NA	ND
Carbamazepine	4/17	3.80 $\pm$ 4.38	8/33	6.33 $\pm$ 3.79
Nerve blocks	3/0	6.75 $\pm$ 4.76	5/0	10.00 $\pm$ 0.0
Hypnosis	2/3	5.00 $\pm$ 4.24	0/NA	ND

## **Treatment of pain:**

- None of the medications has shown to provide complete pain relief
- Even patients who were on opioids reported only partial pain relief
- Most patients were on a combination of analgesics (pain medications) and some sort of therapy
- Interestingly in this study chiropractic manipulation provided the greatest relief although only 4 patients reported using it
- Patients undergoing massage had more relief than using aspirin or ibuprofen

## Treatment of Cramps and fasciculation:

- Traditional treatments showed some benefit with medications like gabapentin, pregabalin
- Sometimes addition of baclofen or tizanidine might be helpful
- A Cochrane review found that there is not enough evidence to say that medications prescribed for cramps are beneficial
- Mexiletine was recently shown to be beneficial for cramps and fasciculation in patients with ALS

## General recommendations on managing pain:

I prefer a holistic approach with a combination of medications, exercise/stretching and braces. They include:

- pool therapy in warm water
- myofascial release (a specialized stretching technique)
- Massage
- Bracing where needed
- maximal use of adaptive devices, such as lifts, beds, cushions and power chairs (without waiting until the patient falls and breaks a hip to prescribe a chair)
- controlling weight and optimizing nutrition
- detection and treatment of depression

**Questions?**



## Acknowledgements:

1. Our patients and family
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A large, dimly lit cave with a prominent waterfall cascading from a high opening in the ceiling. The cave walls are covered in stalactites and other rock formations. In the foreground, a large crowd of people is gathered on a paved area, looking towards the waterfall. To the right, a large golden Buddha statue is visible. In the background, several white stupas or pagodas are illuminated. The overall atmosphere is one of a significant religious or historical site.

**THANK YOU**