Phenomenology of Leg Stereotypy Syndrome

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BACKGROUND

Leg stereotypy syndrome (LSS), previously named leg stereotypy disorder [1], consists of repetitive, rhythmic, stereotypic leg movements, primarily when seated but occasionally also when standing.

Although a common and familial condition that must be differentiated from other leg movements, including restless legs syndrome, akathisia, and tics, LSS has not been studied in a systematic fashion.

The prevalence of LSS is approximately 7% in a randomly selected healthy population and 17% in a cohort consisting of patients with mixed movement disorders [2].

OBJECTIVE

To describe the phenomenology of leg stereotypy syndrome.

METHODS

We reviewed our video database of patients seen at the Baylor College of Medicine Parkinson's Disease Center and Movement Disorders Clinic to identify patients with LSS according to published criteria [1,2].

A detailed review of each video was then conducted in order to characterize the phenomenology.

All patients were subsequently administered a questionnaire capturing clinical questions of relevance to LSS.

Diagnostic research criteria for leg stereotypy syndrome [2]:

- Involuntary, repetitive, and continuous legs moving while seated.
- No diurnal variation in movements.
- Movements occur typically during concentration.
- Movements may be associated with an inner feeling of restlessness.
- Leg movements were self-aware or noted by others.

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RESULTS

- We identified 20 patients with a diagnosis of LSS and video documentation.
- The mean age (SD) at onset of symptoms was 11.3 years (7.9) and the mean age at the time of video recording was 37.7 years (21.1) with a mean duration of symptoms of 26.3 years (17.7). No one reported diurnal variation.
- The majority (17 or 85%) of patients reported onset in childhood (<18 years), all were aware of their movements, and most (17 or 85%) reported experiencing an inner sense of restless or urge, although this was not the main driver of their leg movements.
- Fourteen patients (70%) reported having at least one first degree family member with similar leg shaking.
- On a 5-item LSS severity rating scale, symptoms were generally mild and not associated with adverse outcome with home or work activities.
- On review of the videos, LSS was predominantly unilateral in five (25%) and bilateral in 15 (75%). All subjects had symptoms when sitting: 18 (90%) had rhythmic, flexion-extension of the hip, knee, and ankle, six (30%) exhibited side-to-side movements of the legs, and seven (35%) exhibited inversion-eversion of the ankle. No patients had symptoms only when standing. Of the seven (35%) patients who also had symptoms while standing, four (20%) had leg flexion or knee buckling and five (25%) displayed swaying movements of the body.

Demographics	N	%
Male gender	14	(70)
Ethnicity		
- White	14	(70)
- Hispanic	4	(20)
- Asian	1	(5)
- Black	1	(5)
Age at visit, Mean (SD)	37.7	(21.1)
Age at symptom onset, Mean (SD)	11.3	(7.9)
Duration of LSS in years, Mean (SD)	26.3	(17.7)
Most common co-morbidities		
- ADHD	11	(55)
- TS	10	(50)
- OCD	8	(40)
- Depression	4	(20)
- RBD	4	(20)

Video Review	N	%
Laterality		
- Right leg	3	(15)
- Left leg	2	(10)
- Both legs	15	(75)
Body position		
- Sitting only	13	(65)
- Standing only	0	(0)
- Sitting and standing	7	(35)
Sitting		
- Flexion/extension [ankle, knee, and hip]	18	(90)
- Abduction/adduction [hip]	6	(30)
- Inversion/eversion [ankle]	7	(35)
Standing		
- Flexion/extension [ankle, knee, and hip]	4	(20)
- Swaying of the body	5	(25)

Symptomatology	N	%
Timing		
- Anytime	20	(100)
- Evening only	0	(0)
Distressing		
- To self	0	(0)
- To others	12	(60)
Worsening factors		
- Concentration	12	(60)
- Boredom	15	(75)
- Other	9	(45)
Associated with inner sense of restlessness or urge	17	(85)
Pleasant sensation associated with LSS [e.g., comfort, soothing]	19	(95)
Suppressible		
- Yes	16	(80)
- No	4	(20)
Family history		
- None	6	(30)
- 1 relative	10	(50)
- >1 relative	4	(20)
Treatment		
- Interested in treatment	4	(20)
- Previously tried treatment	0	(0)

Severity Rating Scale*	Mean	SD
Overall severity	1.6	(0.9)
Need or urge to shake legs	2.1	(1.1)
Frequency of leg shaking	2.3	(0.9)
Impact on activities at home and work	0.2	(0.5)
Change during the past year	1.1	(0.7)

*0-4 Likert scale (0 = no symptoms or never, 4 = severe or constant)

MODIFIED LEG STEREOTYPY SCREENING QUESTIONNAIRE

1. Have you ever noticed your legs moving involuntarily, repetitively and continuously while sitting in a meeting, waiting rooms, car, airplane or other situations requiring prolonged sitting?
2. Are the movements associated with an inner feeling of restlessness or urge to move?
3. Do you try to consciously control or suppress the leg movements?
4. Do the leg movements occur any particular time of the day (not just in the evening)?
5. Are the leg movements more likely to occur when you are concentrating?
6. Are the leg movements more likely to occur when you are bored?
7. Are the leg movements distressing, disrupting, or annoying to others?
8. Do you have "swaying" or other involuntary movements of the body when sitting or standing?
9. Do you have any close relatives with similar leg movements?

CONCLUSIONS

LSS is a common, yet poorly characterized, distinct, familial genetic, condition manifested chiefly by repetitive leg and foot movements when sitting or body swaying when standing.

Although the pathophysiology is not well understood genetic factors probably play an important role.

Treatment of LSS is usually not necessary as patients are commonly not bothered by their leg movements, although they may be disruptive to others.

REFERENCES

1. Jankovic J. Leg stereotypy disorder. J Neurol Neurosurg Psychiatry. 2016;87:220–1.
2. Lotia M, York MK, Strutt AM, Jankovic J. Leg stereotypy syndrome: phenomenology and prevalence. J Neurol Neurosurg Psychiatry. 2018. (epub)