

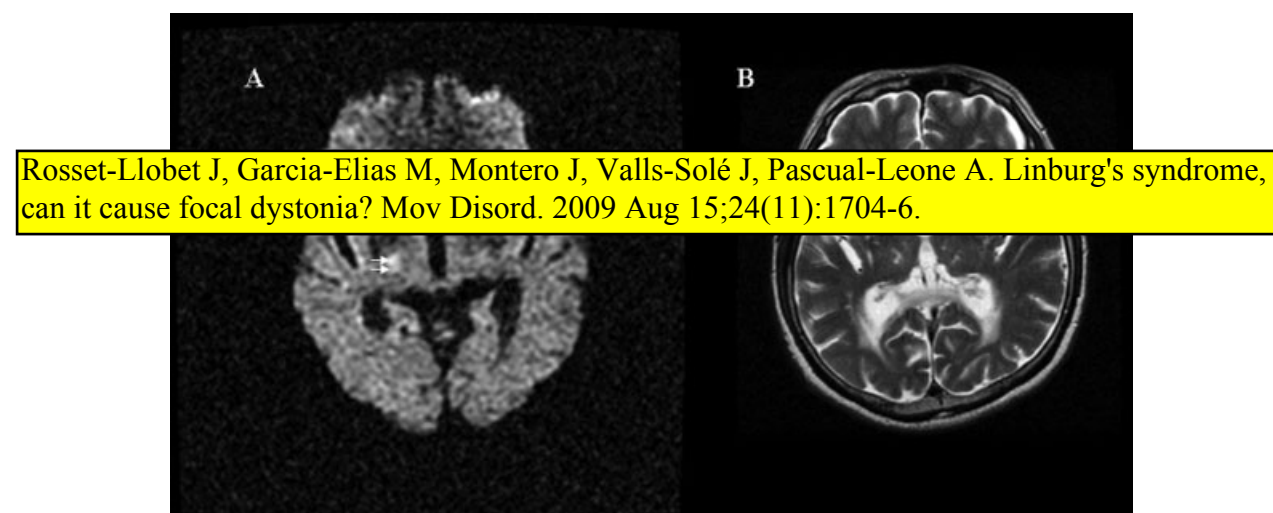


FIG. 1. (A) Initial diffusion-weighted brain magnetic resonance imaging (MRI) scan obtained 14 days after the onset of symptoms. Note the two contiguous lesions with different high signal intensities in the posterolateral part of the right thalamus (arrows). (B) T2-weighted brain MRI showing multiple old ischemic lesions in the bilateral thalami and basal ganglia.

The present case showed that isolated lingual dystonia can be caused by thalamic infarction. Therefore, thalamic lesions should be considered in cases of acute onset lingual dystonia.

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Linburg's Syndrome, Can It Cause Focal Dystonia?

Video 

Repeating movements of high complexity changes the functional organization of the central motor nervous system. In the case of musicians, such plasticity can be maladaptive, and cause task-specific focal dystonia,¹ which may be triggered by biomechanical constraints.²

In the Linburg-Comstock syndrome,³ an aberrant connection between the tendons of the thumb (flexor pollicis longus, FPL) and the index finger (flexor digitorum profundus, FDP), limits active flexion of the thumb interphalangeal joint if there is no simultaneous flexion of the index' distal interphalangeal joint. This constraint is present in a third of the general population,⁴ but is not known to generate specific problems among musicians.⁵

A 32-year-old male classical pianist presented with an involuntary flexion of the index finger while playing piano, which he noticed during a concert 6 weeks earlier. He reported always experiencing some constrain moving the thumb with his index finger extended, but did not recall any musical pieces where this movement was repeated intensively or where he could not reconfigure the fingering of the passage. However, for the preceding 2 months he had been practicing Chopin OP 25-11, which requires repeated movements of the thumb to various keys while the index finger is extended. He experienced some difficulties during the preparation of the concert but denied involuntary

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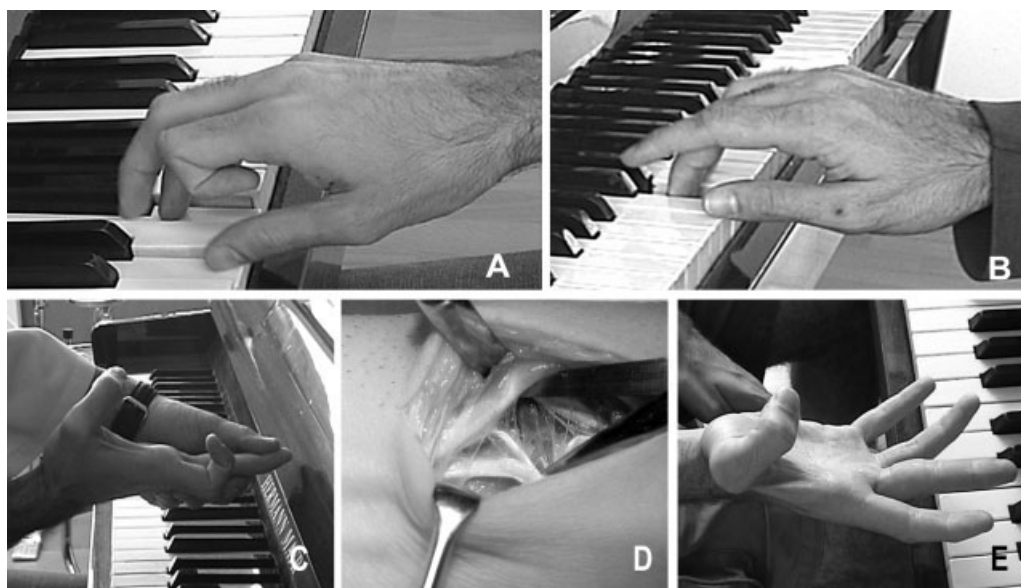


FIG. 1. In this musician, dystonia is seen as involuntary flexion of the index finger (A). Six months after the onset of the reeducation program, the involuntary index finger flexion is no longer seen playing on the piano or other activities (B). Before surgery, thumb flexion is not possible without simultaneous index finger flexion (C). Surgical inspection of the tendons demonstrates a connection between FPL and FDP tendons (D). After surgical release of the connection, thumb flexion is possible without simultaneous index finger flexion (E).

movements or contractions till the concert day when he first noticed the problem.

Physical exam revealed an apparently forced flexion of the index finger that appeared only during piano playing (Fig. 1A), and the impossibility to maintain flexion of the thumb without accompanying flexion of the distal interphalangeal joint of the index finger (1C). Neurological examination, nerve conduction test, and electromyogram did not reveal other abnormalities. Our working diagnosis was task-specific focal dystonia in a patient with Linburg-Comstock syndrome.

After surgical release of the tendons (Fig. 1D), the patient was able to perform independent distal interphalangeal joint flexion of the index and the thumb (Fig. 1E). He began cautiously to practice piano 4 days after the operation, and reported a significant, albeit not complete, improvement in his performance. Unfortunately, the improvement remitted after a few days, and he began to complain again of involuntary flexion of the index finger. We initiated a neuro-rehabilitation program based on Sensory Motor Retuning⁶ that involved daily sessions of work on the piano performing seven different digit combinations while some of the fingers splinted for ~90 min, followed by practice of slow and easy movements without splints. Every month, the practice schedule, specific exercises, kind of digit combinations, and digit positions during the work were checked and adjusted if needed. The goal is to promote alternative motor programs based on modification of the proprioceptive input. The patient showed a progressive improvement of his condition, and task-induced involuntary flexion of the index finger was gone after 6 months (Fig. 1B). Eventually, he returned to his professional activity and his symptoms have not returned over the past 2 years.

The Linburg-Comstock syndrome is a congenital condition and, therefore, our patient certainly had the mechanical constraints related to it all his life. Likely, he was well adapted to this condition, with no apparent motor difficulties until

playing the Chopin concert. As this piece does not allow alternative movements, he was forced to train in adverse biomechanical conditions, which might have led to the development of unwanted motor synergies and ultimately the manifestation of focal dystonia.

After surgical repair of the Linburg-Comstock syndrome, biomechanical, cutaneous, and proprioceptive changes appear to have been sufficient for a transient improvement of our patient's dystonia. In some musicians, focal dystonia can be relieved by postural or mechanical strategies, like having a piece of tape attached to one finger, or holding an object between two fingers. Such "tricks" might work because changes in sensory inputs may improve selection of motor commands⁷ or minimize abnormalities of sensory processing.⁸ Unfortunately, dystonic movements often reappear after a variable time of practice (minutes to days). Similarly, the post-surgical benefit in our patient was time-limited but further recovery was possible with Sensory Motor Retuning, which might have induced a more consistent change in the dysfunctional central nervous system circuits.⁶

Our case reveals that, even when an apparent precipitating event is clearly recognized, dystonia, which involves a plastic change in the central nervous system, may remain after the apparent precipitating cause has been removed. The process of re-learning the specific motor task, such as with Sensory Motor Retuning, may succeed in improving motor control. However, we speculate that, in the particular case reported here, surgical intervention could have been partially useful by facilitating re-education under more favorable biomechanical conditions.

LEGENDS TO THE VIDEO

Segment 1. Flexion of the interphalangeal joint of the thumb is not possible without index finger flexion. The

patient was requested to flex the thumb but this was not possible to do in isolation (without flexing the index finger too), due to an anomalous connection between the tendons of the two muscles.

Segment 2. Involuntary index finger flexion and middle finger extension. After practicing piano playing and trying to perform movements requiring simultaneous thumb flexion and index extension, the patient began to feel involuntary movements on the index and middle finger.

Segment 3. Post-surgery: flexion of the interphalangeal joint is now possible without simultaneous index flexion. Surgical liberation of the anomalous tendinous connections allowed isolated thumb movements.

Segment 4. Reduction of involuntary movements. Surgical liberation of the tendons led to a clear amelioration of motor performance, with less unwanted index finger flexion and middle finger extension.

Segment 5. Involuntary movements reappeared after 15 days of practicing piano. The patient was asked to slowly return to piano practice after surgery. However, the involuntary index finger flexion and middle finger extension reappear 2 weeks after starting practice.

Segment 6. Involuntary movements improved after 6 months of reeducation. Guided repetitive daily exercises on the piano led to a significant improvement of finger control, with a consistent reduction in index finger flexion and middle finger extension.

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“Acquired” Hepatocerebral Degeneration in a Patient Heterozygote Carrier for a Novel Mutation in *ATP7B* Gene

Acquired hepatocerebral degeneration (AHD) is a rare type of hepatic encephalopathy characterized by neuropsychiatric symptomatology, and peculiar neuroradiologic findings, without the clinical evidence of Wilson's disease (WD).^{1,2} We studied a patient with AHD responsive to penicillamine who was heterozygote carrier for a novel mutation in the *ATP7B* gene, and discussed the possible role of the mutation in facilitating the appearance of the syndrome.

A 37-year-old man with liver cirrhosis related to chronic hepatitis C was admitted because of progressive consciousness impairment. Family history was negative for WD. Ammonemia was 176 $\mu\text{mol/L}$ (NR, 9–33 $\mu\text{mol/L}$); total bilirubin 1.71 mg/dL (NR, 0.2–1.3 mg/dL); serum albumin 2.6 g/dL (NR, 3.3–5 g/dL); AST 62 U/L (NR, 10–45 U/L). Electroencephalogram disclosed diffuse slow wave activity. After rifaximin, lactulose, and branched chain amino acid infusion, his arousal state went back normal in about 12 hours, and ammonia levels decreased to 94 $\mu\text{mol/L}$.

Neurological examination revealed bradykinesia, sialorrhea, dysarthria, mild rest tremor, in the upper limbs, prevalent on the right side, postural instability, and camptocormia. Moreover, he showed depression and anxiety. The MMSE had normal scores (27/30).

Serum ceruloplasmin, cupremia, and cupruria were within the reference values. Ocular slit-lamp examination did not reveal Kayser-Fleischer ring. CSF examination revealed increased manganese concentration (2.9 $\mu\text{g/L}$; NR, 0.88 $\pm$ 0.76 $\mu\text{g/L}$), while copper and iron levels were within normal values. Brain MRI showed increased signal intensity in basal ganglia bilaterally on T1-weighted images (Fig. 1A,B). Levodopa and pramipexole were of no benefit. After informed consent, he was treated with penicillamine 900 mg daily and trihexyphenidyl 2 mg twice daily. In the following 5 days, psychiatric disturbances progressively abated, while bradykinesia,

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