Challenges in Managing Dystonia

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International Federation of
CLINICAL NEUROPHYSIOLOGY

International Parkinson and
Movement Disorder Society

Challenging Case #1

- 52-year-old right-handed woman
- Onset: L hand tremor ~2012
- Parkinson’s disease complicated by motor fluctuations
- Severe dystonia of both feet in the OFF state
- Bothersome dyskinesia in the ON state

Current Parkinson's disease medications:

	~0400	~0600	~0800	~1000	~1200	~1400	~1600	~1800	2100
Carbidopa/levodopa IR 25/100 mg	1 tab	1 tab	1 tab	1 tab	0.75-1 tab	0.75-1 tab	0.75-1 tab	0.75-1 tab	
Carbidopa/levodopa CR 25/100 mg									2 tabs
Pramipexole ER	0.375 mg								0.375 mg
Selegiline	5 mg								5 mg

OFF/ON evaluation

- OFF MDS-UPDRS III = 30
- L>R dystonic posturing of the feet with gait impairment
- L>R tremor

- ON MDS-UPDRS III = 5
- Generalized dyskinesia

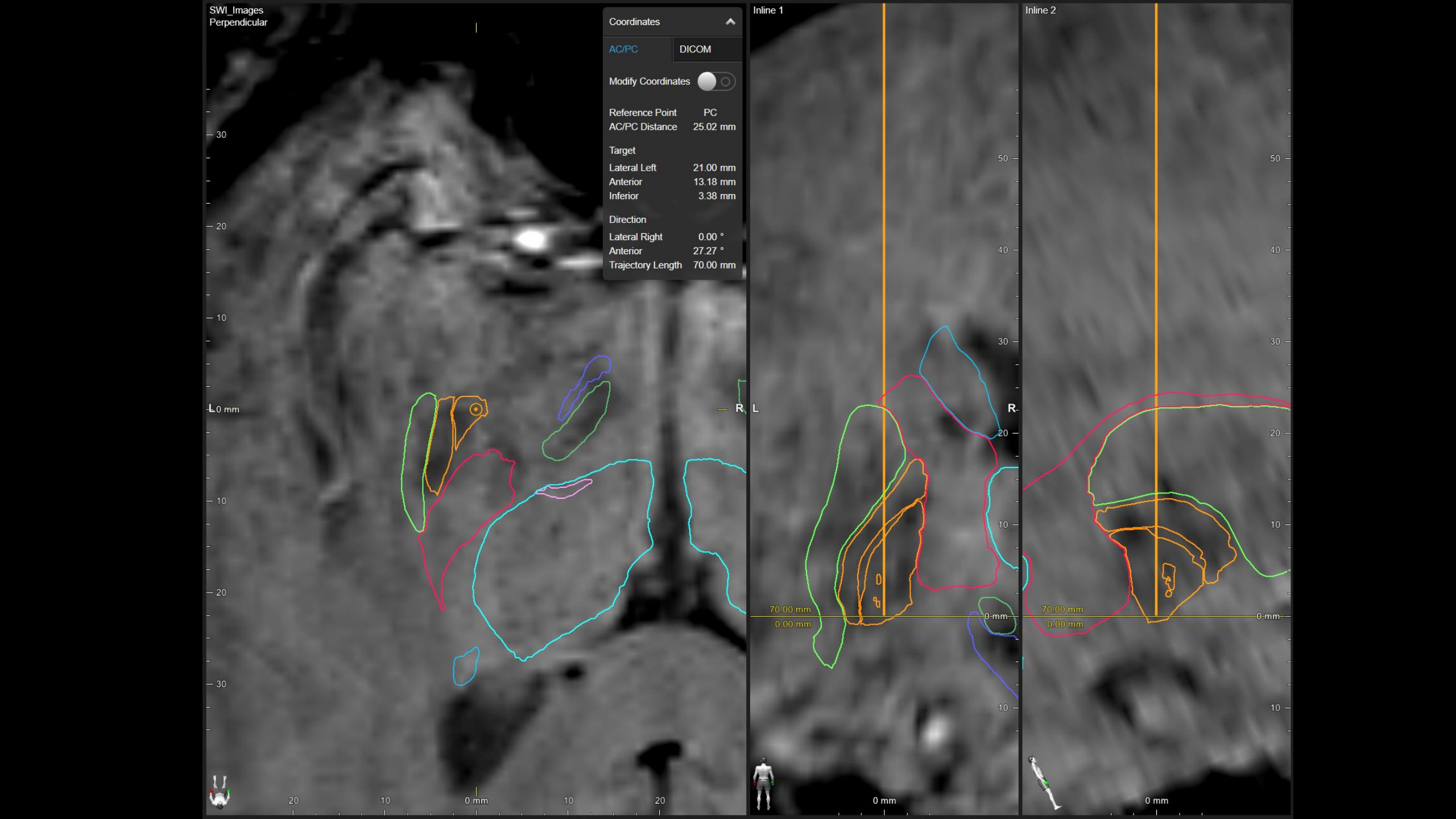
- OFF → ON = 83.3%

DBS candidate?

Pre-operative assessments

- Neuropsychological testing OK
- MRI brain OK
- Neurosurgery OK

- **Target?**

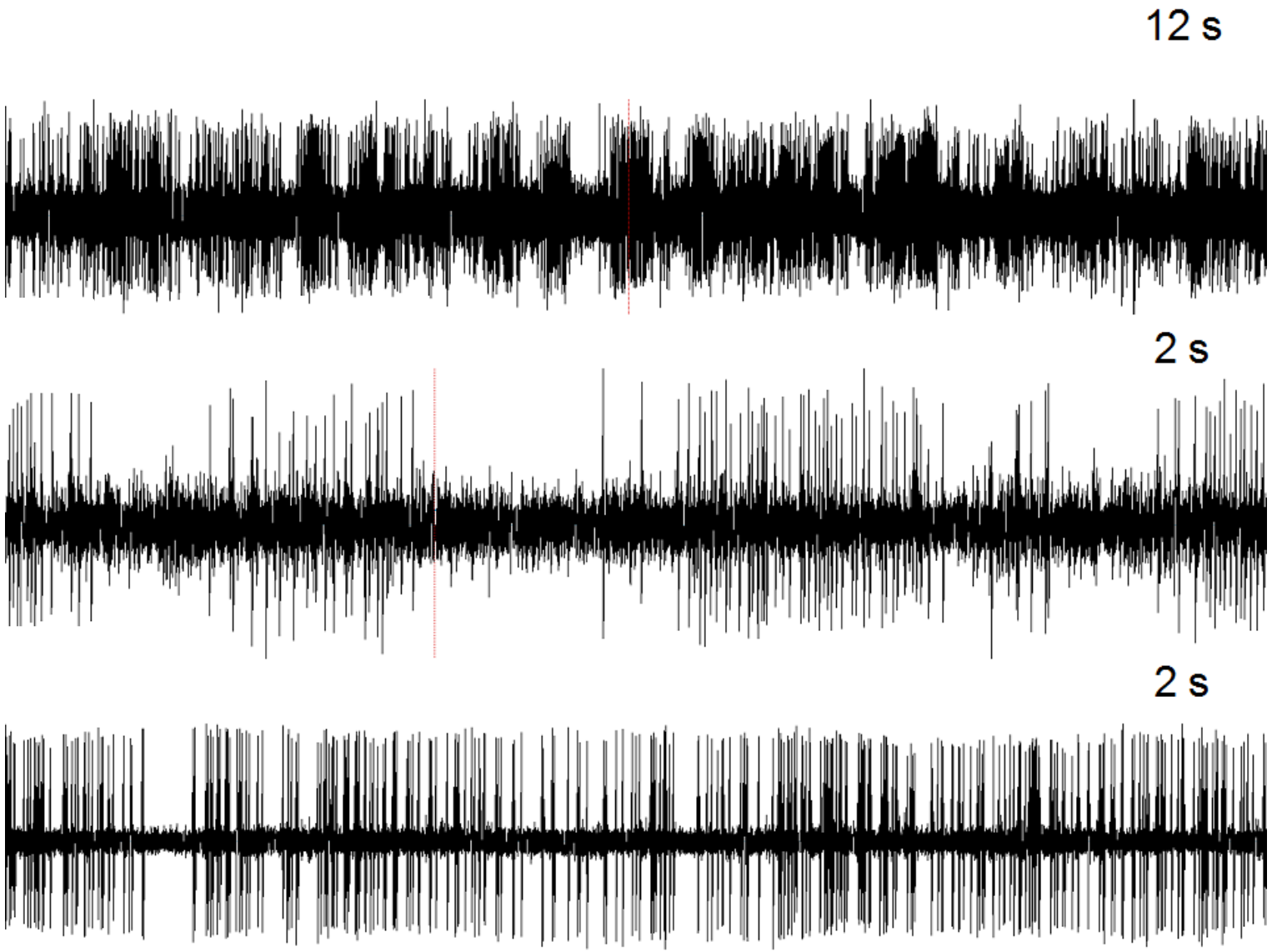

SWI_Images
Perpendicular
Coordinates
AC/PC
DICOM
Modify Coordinates
Reference Point PC
AC/PC Distance 25.02 mm
Target
Lateral Left 21.00 mm
Anterior 13.18 mm
Inferior 3.38 mm
Direction
Lateral Right 0.00 °
Anterior 27.27 °
Trajectory Length 70.00 mm
Inline 1
Inline 2
70.00 mm
0.00 mm
0 mm

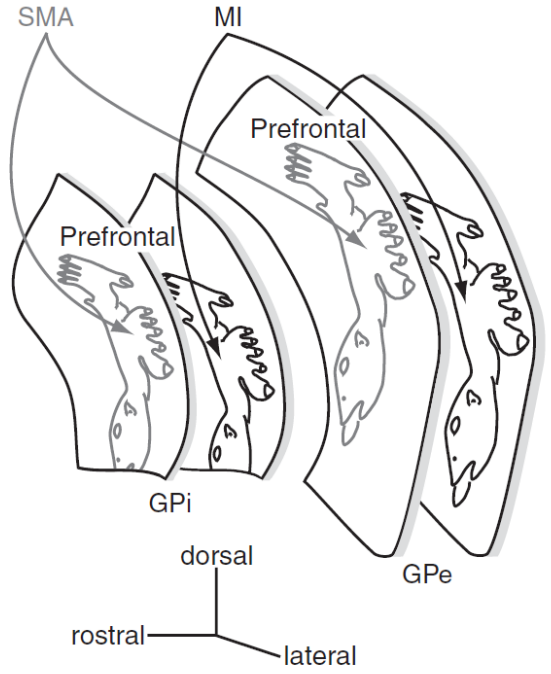
Intra-operative neurophysiology

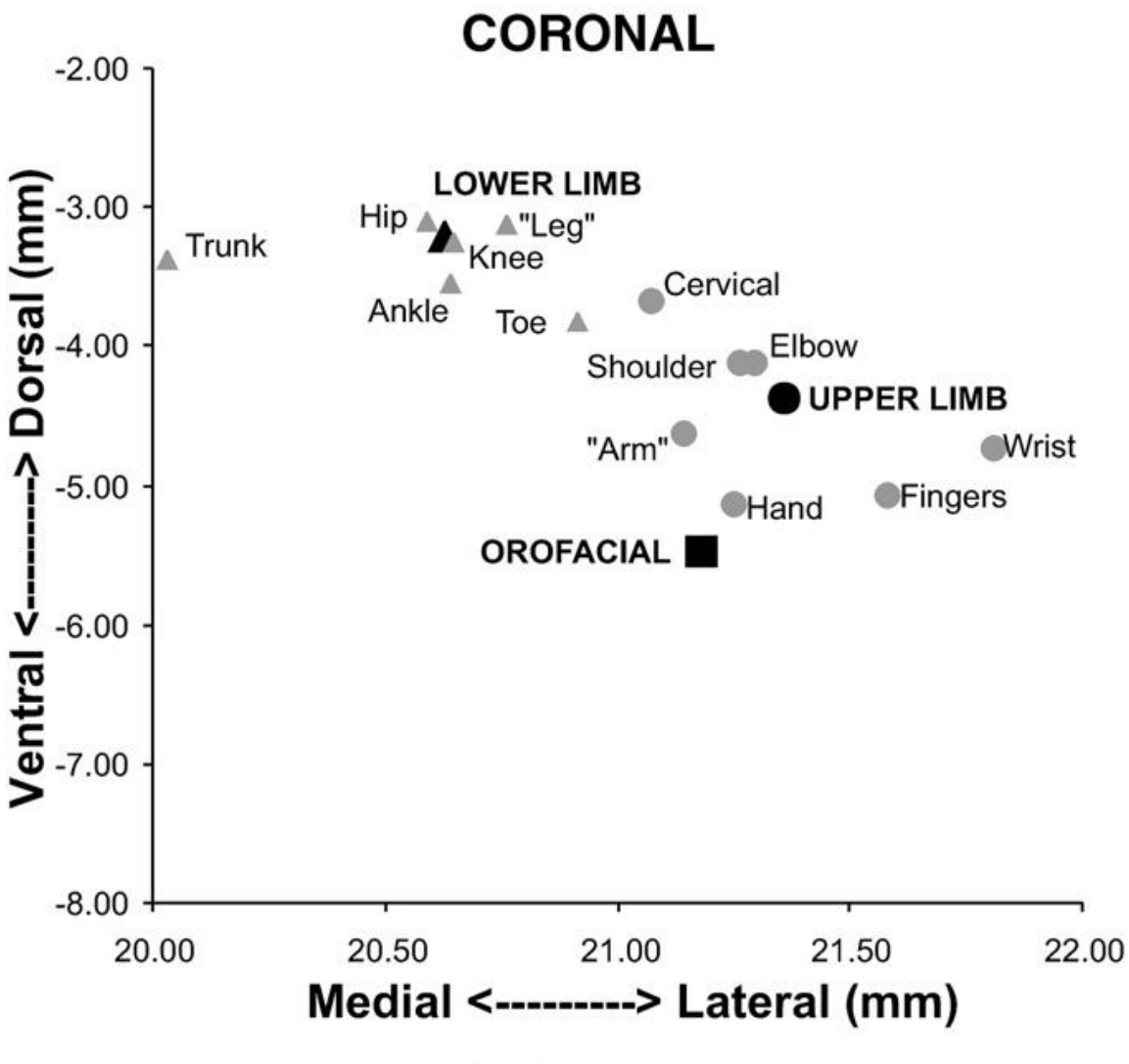
- Microelectrode recordings: single neuron potentials, LFPs
- Microstimulation
- Macrostimulation
- DBS lead: Stimulation testing, LFP measurement

Table 1. Common neuronal firing patterns and stimulation-induced effects encountered during deep brain stimulation surgery for movement disorders.

Structure	Neuronal firing pattern	Stimulation-induced effects
SNr	High frequency (regular), moderate density, background and amplitude	Mood changes, worsening of Parkinsonism
GPe (more evident in PD than in dystonia)	Moderate background. Brief interruptions of moderate frequency, continuous activity (pausing cells). Brief bursts of high frequency activity (bursting cells)	Dyskinesia
GPi	High density, amplitude and frequency, low to moderate background. Bursting cells that may correlate with tremor. Sensorimotor activity	Dyskinesia (dorsal GPi), improvement of Parkinsonism (in PD)
Medial lemniscus	Absent or markedly reduced (white matter)	Contralateral paresthesia
Internal capsule	Absent or markedly reduced (white matter)	Contralateral muscle pulling, twitching, posturing, gaze deviation, dysarthria
Red nucleus	Similar to STN	Nausea, sweating, warm/cold sensation, dizziness, etc.
Oculomotor nucleus	NA (Not part of trajectory or target)	Ipsilateral esotropia or esophoria (with or without diplopia)
Optic tract	Absent or markedly reduced (white matter). May be evoked by light stimulus	Contralateral phosphenes


12 s
2 s
2 s





Baker KB, Lee JY, Mavinkurve G, Russo GS, Walter B, DeLong MR, Bakay RA, Vitek JL. Somatotopic organization in the internal segment of the globus pallidus in Parkinson's disease. Exp Neurol. 2010 Apr;222(2):219-25. doi: 10.1016/j.expneurol.2009.12.030. PMID: 20059997

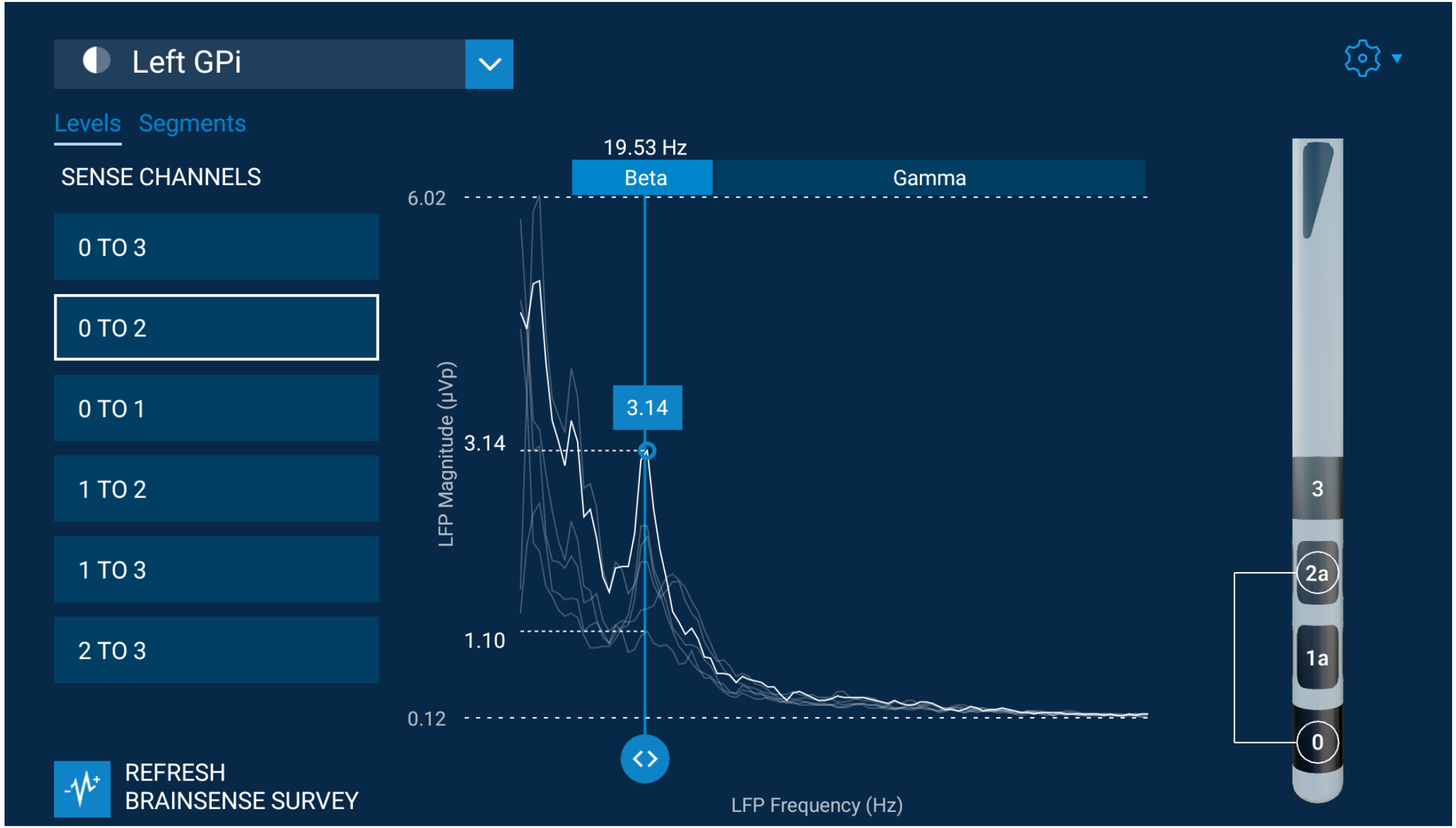
Table 2. Local field potentials associated with movement disorders.

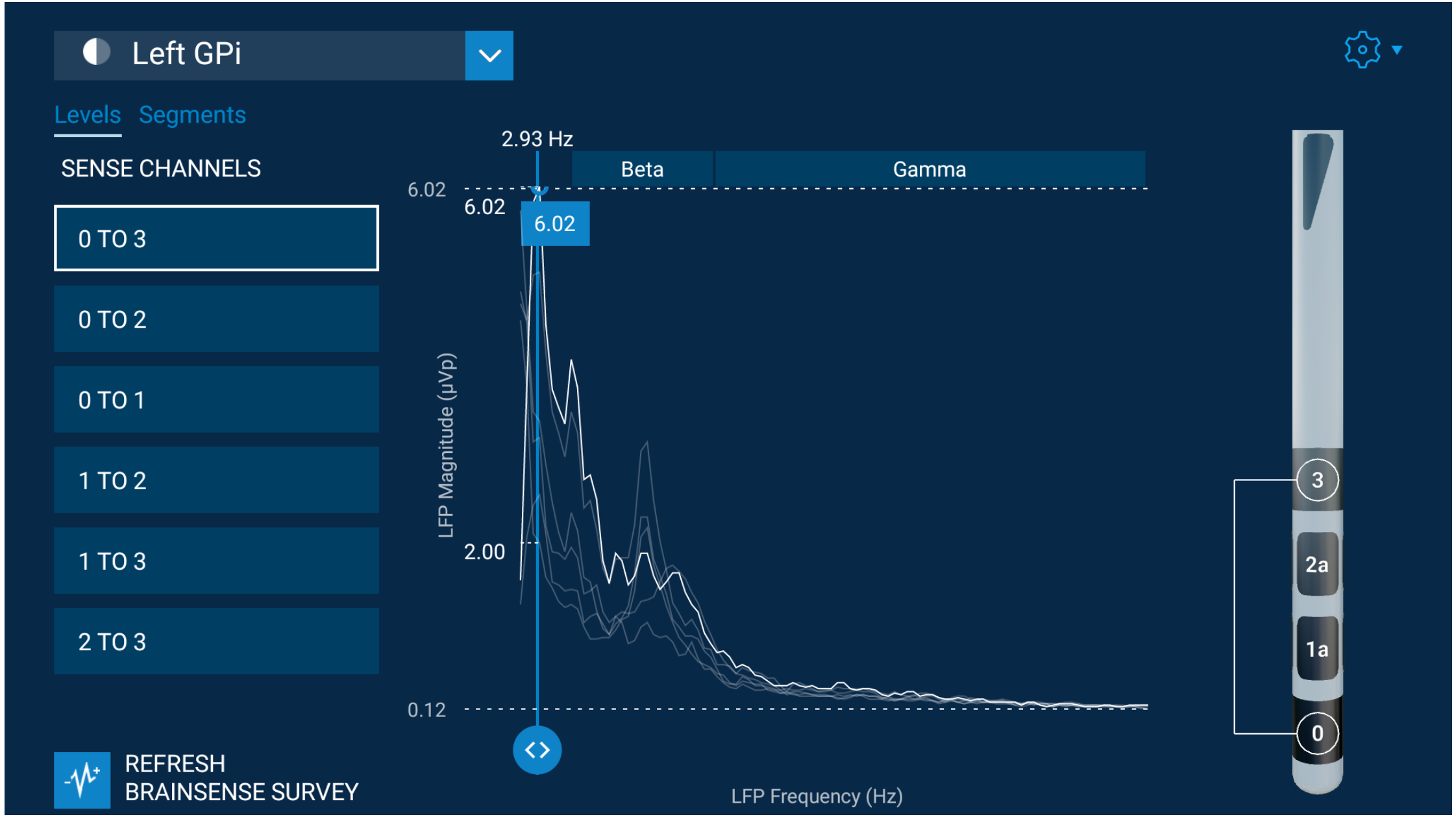
Movement disorder	Local field potentials (frequency peak)	
	Increased synchronization	Reduced synchronization
Essential tremor	■ Theta/alpha (4–13 Hz) ■ Beta† (13–35 Hz)	
Levodopa-associated dyskinesia	■ Theta/alpha (4–13 Hz) ■ Gamma† (60–90 Hz) (coherent between STN and motor cortex)	■ Beta† (13–35 Hz)
Dystonia (tonic)	■ Delta (0–4 Hz) ■ Low beta (13–20 Hz) (shorter bursts) ■ Gamma (60–90 Hz)	
Dystonia (phasic)	■ Theta/alpha (4–13 Hz) ■ Low beta (13–20 Hz) (shorter bursts) ■ Gamma (60–90 Hz)	
Tics	■ Theta/alpha (4–13 Hz)	

†Also associated with initiation and facilitation of normal voluntary movements.

L GPi

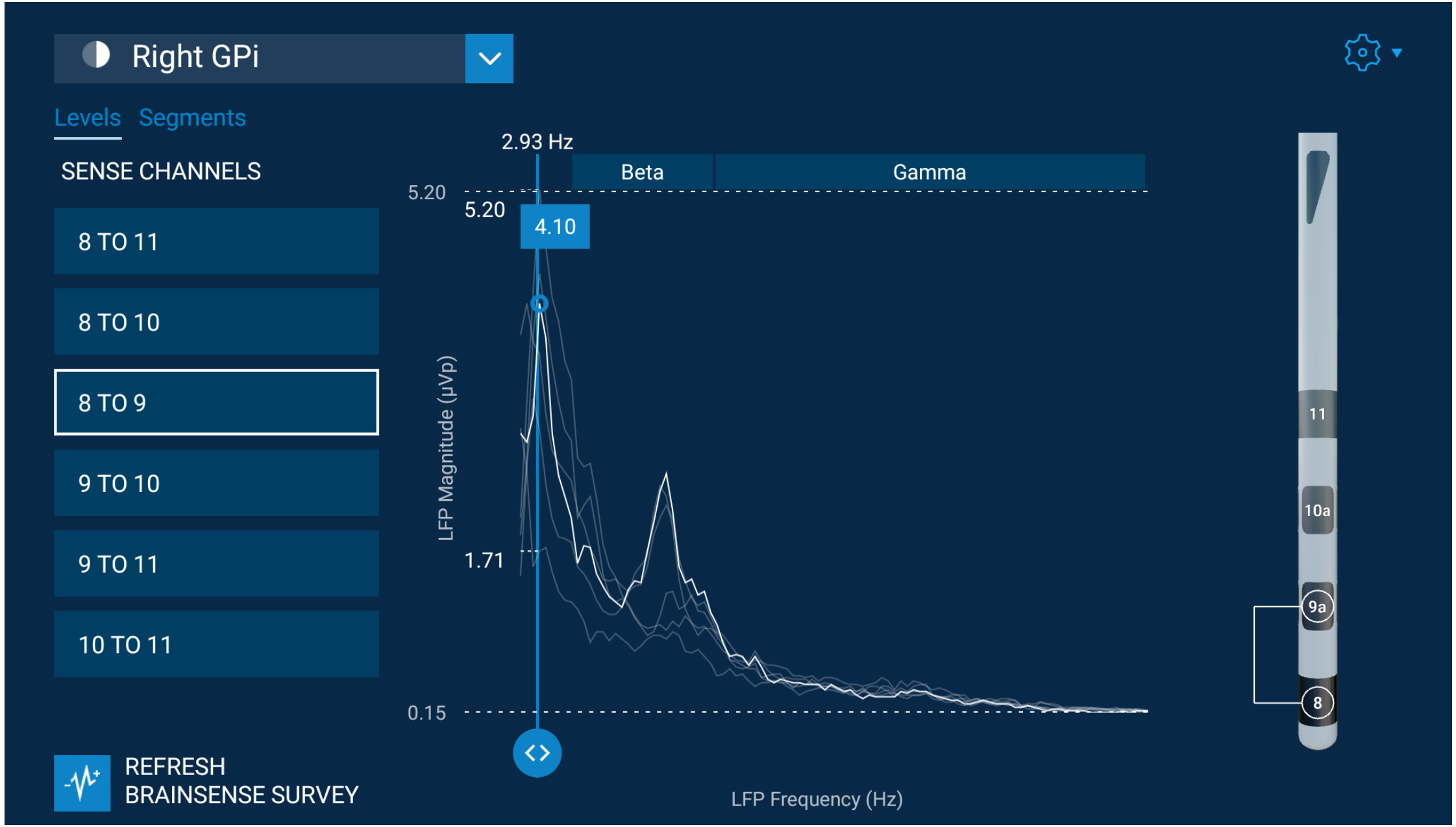
OFF


Left GPi
Levels
Segments
SENSE CHANNELS
0 TO 3
0 TO 2
0 TO 1
1 TO 2
1 TO 3
2 TO 3
REFRESH
BRAINSENSE SURVEY
19.53 Hz
Beta
Gamma
6.02
3.14
1.10
0.12
LFP Magnitude (µVp)
LFP Frequency (Hz)
3
2a
1a
0


Left GPi
Levels
Segments
SENSE CHANNELS
0 TO 3
0 TO 2
0 TO 1
1 TO 2
1 TO 3
2 TO 3
REFRESH
BRAINSENSE SURVEY
2.93 Hz
Beta
Gamma
6.02
6.02
6.02
2.00
0.12
LFP Magnitude (µVp)
LFP Frequency (Hz)
3
2a
1a
0

R GPi

OFF


Right GPi
Levels
Segments
SENSE CHANNELS
8 TO 11
8 TO 10
8 TO 9
9 TO 10
9 TO 11
10 TO 11
REFRESH
BRAINSENSE SURVEY
2.93 Hz
Beta
Gamma
5.20
5.20
4.10
LFP Magnitude (µVp)
1.71
0.15
LFP Frequency (Hz)
11
10a
9a
8

Thank you