Understanding the educational needs of neurologists who manage patients with Huntington's disease: a US survey study analyzing both clinician and patient perspectives

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1 Introduction

Huntington's Disease (HD) is a hereditary neurodegenerative disease that manifests in a progression of motor, cognitive, and psychiatric symptoms until a patient's death. The clinical diagnosis is typically made based on family history and confirmed by genetic testing, but ultimately is a clinical diagnosis based on motor disorder symptoms and cognitive examination.

There are many challenges to optimizing care in patients with HD. Due to the myriad of symptoms, a multidisciplinary approach is needed for optimal patient management. Coordinating specialist and primary care physicians, nurses, physical and occupational therapists, speech pathologists, genetic counselors, etc. in the care of a patient is often challenging. Furthermore, there is currently no cure for HD, only management of specific symptoms.

In order to design the most effective interventions for future continuing education, this study was conducted in order to determine the needs and barriers of clinicians managing patients with HD. Taking perspectives of neurologists in HD Centers of Excellence (CoEs), general neurologists, as well as patients/caregivers will provide developers and supporters of education real-world data and establish a baseline for measuring educational effectiveness.

2 Methodology

Development of clinician survey

With an HD expert, a case vignette-based survey was developed to assess current practice, attitudes and barriers of neurologists. The survey was piloted with practicing clinicians before launch.

Patient/caregiver survey and IRB approval

An accompanying survey for patients/caregivers was developed with the HDSA. An independent IRB reviewed and approved the patient portion of the study on January 4, 2017.

Data collection

Data were collected through an online survey platform from neurologists and patients/caregivers from January through April 2017.

Analysis

All questions were arrayed in SPSS Statistics 22 and any open-ended questions were coded. Analyses were conducted to determine gaps in practice and areas of future educational need.

3 Demographics of neurologist sample

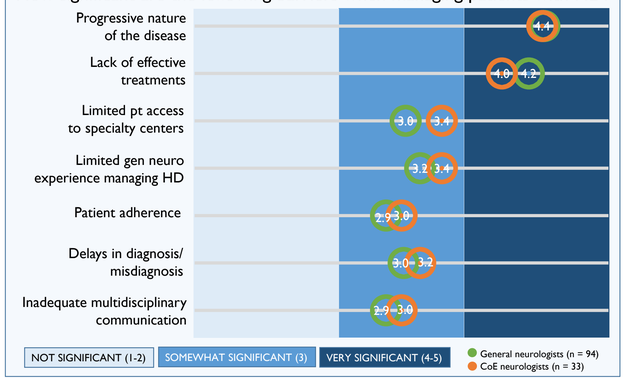
	Center of Excellence (n = 33)	General neuro (n = 94)
Location		
Practice type		
Patients seen each week	45	78
Yearly patients with HD	52	10
Medical school graduation year	2001	1988

4 Demographics of patient sample

	HD patient (n = 33)	Caregiver (n = 94)
Gender		
Seen at HD CoE?		
Age	45	46

5 When managing patients with HD, neurologists are most concerned with its progressive nature and lack of effective treatments.

How significant are the following barriers when managing patients with HD?

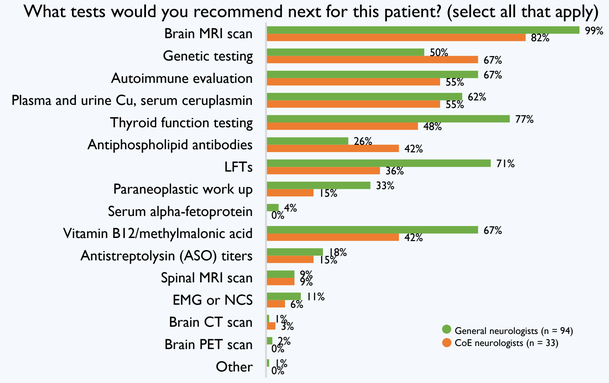


CASE: A 51-year-old man is referred for a 12-month history of gradually progressive balance difficulties and falls. He works as a teacher and has noted increasing difficulty performing his job due to forgetfulness. His wife notes some difficulties with speech, specifically with enunciation. The patient reports that he is more restless and fidgety at times. He otherwise remains completely independent in chores and activities of daily living, though possibly slower. He denies depression or anxiety, and his wife concurs. There is no family history of similar disorder.

On examination, he is oriented without aphasia, recent and remote memory is preserved, and there is no apraxia. He has slight latency on saccade initiation and ocular pursuits are jerky but full range. He has motor impersistence on eye closure and tongue protrusion. He has mild, generalized chorea of the face, trunk, and all four limbs. Voluntary movements are irregular. His gait is wide-based and irregular, and he has difficulty with tandem gait.

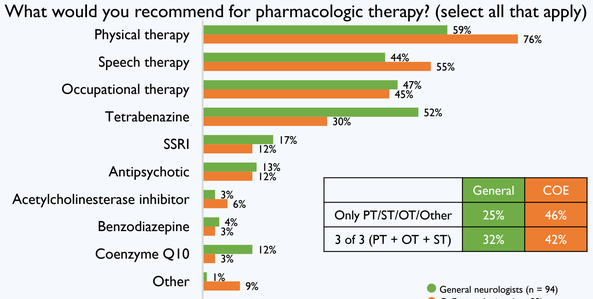
6 Neurologists generally included appropriate tests in their diagnostic workup.

What tests would you recommend next for this patient? (select all that apply)



7 Nonpharmacologic management is preferred at this point but some are choosing other treatments.

What would you recommend for pharmacologic therapy? (select all that apply)



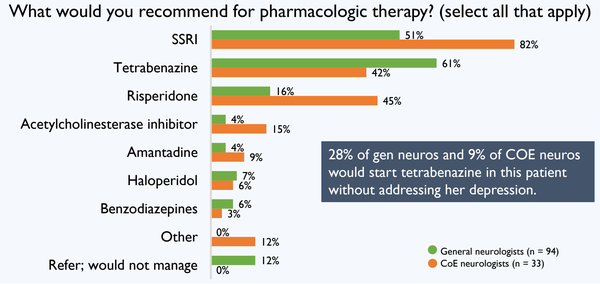
	General	COE
Only PT/ST/OT/Other	25%	46%
3 of 3 (PT + OT + ST)	32%	42%

Case #2: A 47-year-old woman is seen for a possible diagnosis of HD. She notes progressive abnormal jerking movements that started about 1 year ago. She drops things and has near daily falls. She has 2-year history of depression and anxiety that has gotten worse since losing her job, with thoughts of suicide but denies an active plan or intent. She notes that her memory has been problematic, such as missing appointments and bills. Her family history is extensive for HD.

On exam, she has relatively preserved cognitive function with intact recent and remote memory. Her affect is sad and she would become tearful at times. She has head thrusts to initiate saccades and slowing of saccadic eye movements. She has normal tone and full strength, but motor impersistence on tongue protrusion. She has generalized grade 3 chorea on UHDRS examination and a total chorea score of 21. Gait was wide based with choreic intrusions. She was unable to perform tandem gait and would retropulse on pull testing.

8 While many neurologists in CoEs would initially start this patient on an SSRI, some would immediately begin tetrabenazine.

What would you recommend for pharmacologic therapy? (select all that apply)

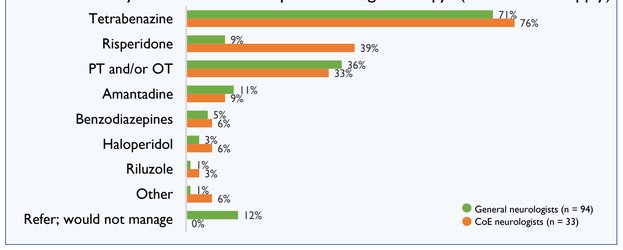


28% of gen neuros and 9% of COE neuros would start tetrabenazine in this patient without addressing her depression.

CASE 2 (cont.): You started her on sertraline 50 mg daily. At her follow-up visit two months later, she reports that her mood is excellent. She is more interactive with family/friends and denies any suicidal ideation. Her involuntary movements remain bothersome, which are interfering with her ability to easily feed and dress herself. The movements are associated with imbalance and occasional falls. On exam, she has moderate-to-severe generalized chorea, grade 2-3, with a total chorea score of 20.

9 After a marked improvement in depressive symptoms with an SSRI, most neurologists would add tetrabenazine.

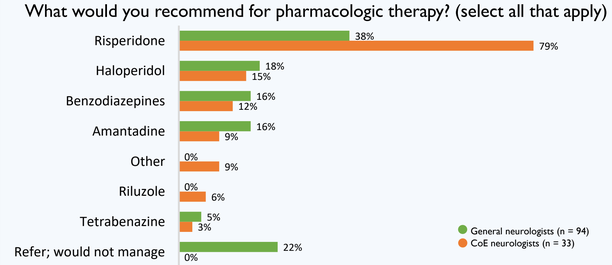
What would you recommend for pharmacologic therapy? (select all that apply)



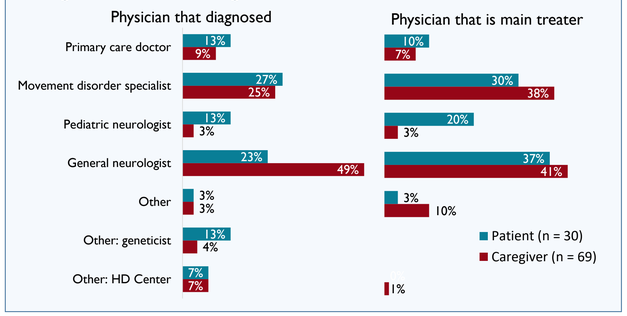
CASE 2 (cont.): The patient is started on tetrabenazine and 4 weeks later, the patient reports feeling more depressed with some suicidal ideation. On exam, she is disheveled, tearful, and bradyphrenic, with a flat affect. You wean her off her tetrabenazine and in 4 weeks her mood is markedly improved, and she denies suicidality. She again has a marked worsening of her chorea with grade 3 chorea of the limbs and trunk and a total chorea score of 18.

10 Neurologists were divided on how to manage a patient's chorea without exacerbating depression.

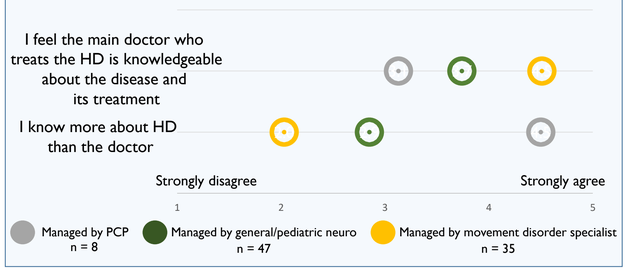
What would you recommend for pharmacologic therapy? (select all that apply)



11 Patients are generally diagnosed and are currently treated by a movement disorder specialist or general neurologist.

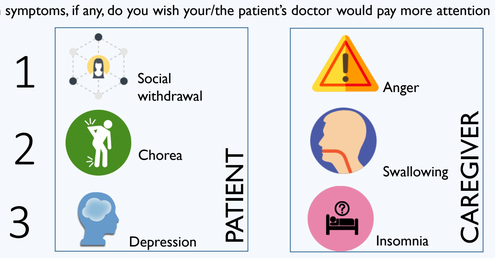


12 Patients and caregivers managed by PCPs feel that their doctor is not as knowledgeable as those managed by a specialist.



13 Patients and caregivers point to different symptoms needing more attention from their physician.

Which symptoms, if any, do you wish your/the patient's doctor would pay more attention to?



14 Discussion

Physicians managing patients with HD, especially general neurologists, may need continued education on key concepts of diagnosis and management. Understanding the relationship between psychiatric/cognitive symptoms and motor symptom treatment is paramount. HCP education should continue to focus on addressing symptoms and providing resources to patients and caregivers on how to manage these symptoms, including tips on daily living and what can be expected in the future.

The right physician is important! Patient and caregiver confidence in their physician's knowledge and competence increased when they were being managed by a specialist. Nearly all of the patients managed by a primary care physician in this study were in urban/suburban areas and not far from HD CoEs.

Caregivers and patients have different educational needs. They prioritize symptoms differently, perceive healthcare providers differently, and have different challenges. The HD community should be used for patient and caregiver outreach, but allow separate breakout programs specifically oriented to these groups.

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