



Huntington's Disease In Service Training for Long Term Care Facilities

Insert date
Your name

Improving the lives of everyone with Huntington's disease & their families.

On behalf of the Huntington's Disease Society of America (HDSA), we thank you for your interest and willingness to care of our patients with Huntington's disease.

Our Mission

To improve the lives of everyone affected by Huntington's disease and their families.

Our Vision

A world free of Huntington's disease.





WHAT WILL WE COVER TODAY?

What is Huntington's disease?

- Clinical features
- HD progression
- Long term care for HD
- Treatment strategies for Huntington's disease
- Common challenges in the care of persons with HD
- Case studies and interventions

Let's Start with a Case Study

Joanna is 50 years old with history of chronic low back pain.

At Age 45: She started complaining of depressive mood, irritability, and lack of pleasure and satisfaction from painting, which was her passion. She was seen by a psychiatrist and started on antidepressants with partial improvement in mood.

At Age 48: She complained of balance problems and falls, which was initially attributed to her back pain. In addition, she noticed abnormal involuntary movements in her arms and feet, especially during moments of significant anxiety.

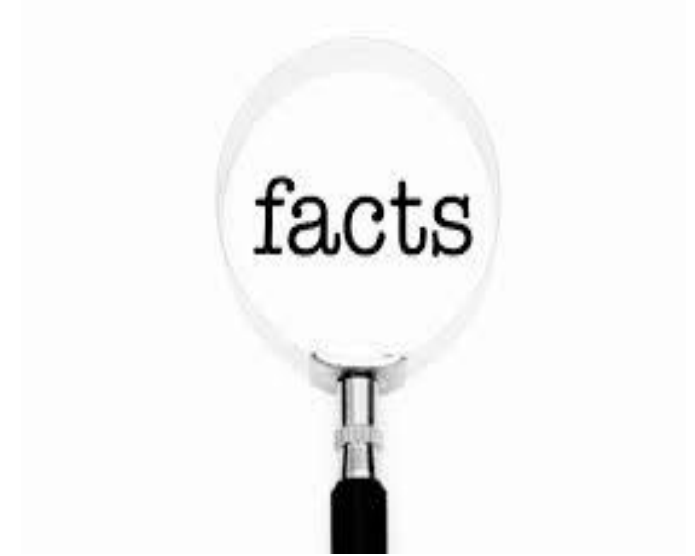
At Age 49: She started having angry outbursts and had a marked increase in the abnormal involuntary movements. Her ability to concentrate at work as supermarket cashier deteriorated and she made multiple mistakes. She was first placed on probation and eventually fired. She attributed her clinical deterioration to the COVID19 pandemic.

Family history: Her father passed away at the age of 78. He had balance problems and abnormal movements of his hands since the age of 53. Even though he was seen by multiple specialists, he never had a clear diagnosis.

Our patient was seen by a neurologist at the age of 50 who, based on the history, was suspicious on Huntington's disease. A genetic test for Huntington's disease was performed, confirming the diagnosis.

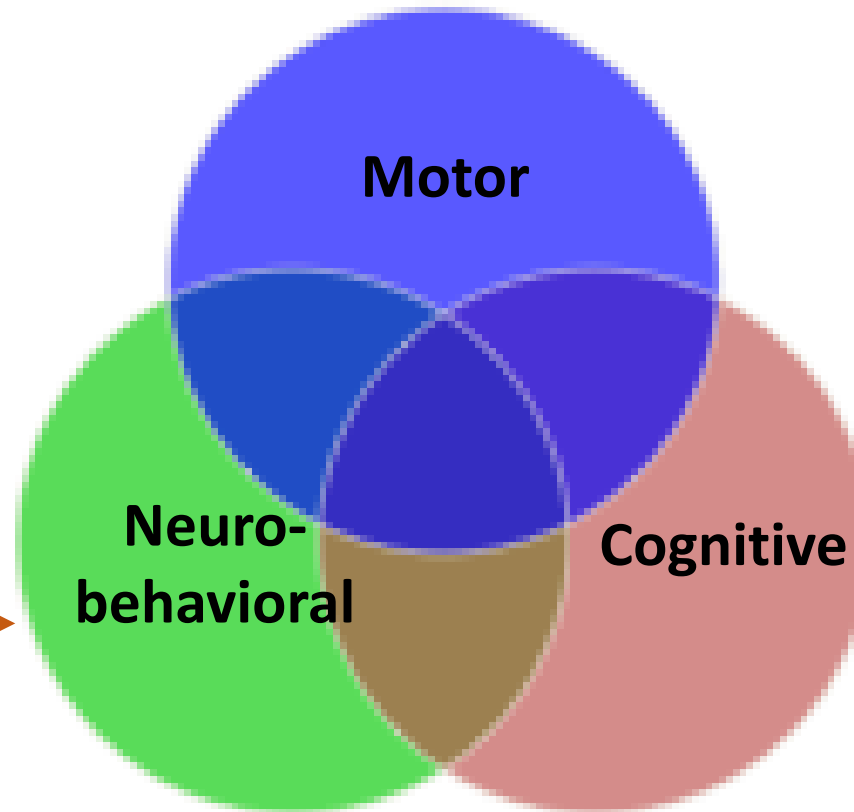
WHAT IS HUNTINGTON'S DISEASE?

- Huntington's disease is a genetic disease.
- Each child of a person with Huntington's disease has a 50/50 chance of developing HD.
- HD symptoms usually start between the ages of 35 to 45. (Range 1-80 years)
- The larger of number of CAG repeats, the earlier HD symptoms can present.
- The mean duration of the disease is 17-20 years.
- There is currently no cure for HD nor any way to slow down the disease progression.
- *However*, there are many symptom management tools and strategies for persons living with HD.



CLINICAL FEATURES OF HD

Disinhibition
Depressed mood
Suicide: 5-10 %
Euphoria
Irritability (possible aggression)
Repetitive thoughts
Delusions (false belief)
Hypersexuality
Hallucinations



Chorea (fast dance-like involuntary movements)
Dystonia (slow involuntary movements causing abnormal postures)
Stiffness/Rigidity
Gait imbalance, falls
Slurred speech
Swallowing difficulties

Executive dysfunction (poor planning, difficulty organizing, poor flexibility to adapt to alternatives, slow thinking process)
Memory loss
Verbal fluency problems
Disorientation, getting lost
Judgement problems
Impaired decision making

WHAT DOES HD PROGRESSION LOOK LIKE?

PHASES OF HUNTINGTON'S DISEASE			
	Stage 1- Early	Stage 2-Middle	Stage 3-Late
Symptoms	Subtle or mild symptoms, manageable with medications	Increasingly intrusive motor, cognitive, or behavioral symptoms	Poorly controlled movements
Location of Care	Home	Home with assistance or assisted living	Skilled nursing care
Activities of Daily Living	Independent	Not driving, managing finances, may need help with self-cares	Fully dependent on others for daily cares
Disability	No disability but may require work modifications	Unable to work	Unable to work
Emotional Cognitive	Depression, suicide risk	Mood changes, irritability, perseveration	Dementia, reduced ability to communicate

Each individual progresses differently with HD; these are general guidelines but aren't necessarily applicable to each person with Huntington's disease.

LONG TERM CARE AND HUNTINGTON'S DISEASE

LTC is a term used to describe medical and non-medical care that is provided over an extended period of time to people who have a chronic illness or disability.

Individuals with HD often need LTC as the disease progresses.

WHY

As the disease progresses and symptoms increase, a person with HD may need more and more assistance for activities of daily living (ADLs). That is when LTC services may be indicated.

GOAL

To help maximize functioning and quality of life when the affected individual is not longer able to live independently.

WHEN

No way to predict. It could be at any age.



INDEPENDENT



DEPENDENT

ROLE OF THE FAMILY IN LONG TERM CARE

AT ADMISSION

- Ask the family about any behavior triggers for the person with HD; the family is the expert on their loved one.
- Ask about who the person was before HD impacted them; what were their interests, career, hobbies.
- Work with the family to manage expectations of care. For example, how often does showering happen, etc.

WHILE IN CARE

- While family involvement is best, HD can create complicated family dynamics and triggers. Family members may be a trigger for a person with HD.
- Don't assume that if family members don't visit that they are uninterested in their person with HD.
- Family can help maintain relationships with HD specialists and teams; utilize them and their expertise.
- Family can be helpful in problem-solving and de-escalation strategies.

OVERVIEW OF TREATMENT FOR HD

MOTOR

- Abnormal movements - chorea: Tetrabenazine (Xenazine), deutetetrabenazine (Austedo), antipsychotics
- Poor balance: Physical therapy, walking aids
- Slurred speech: Speech therapy
- Swallowing problems: Swallowing evaluation and therapy (feeding tube in advanced cases), nutritionist

NEUROBEHAVIORAL (EMOTIONS)

- Depression: Antidepressants, psychiatry/psychology evaluation
- Anxiety: Anti-anxiety agents, psychiatry /psychology evaluation
- Psychosis/hallucinations: anti-psychotics
- Irritability / aggressiveness: tranquilizers (anti-anxiety medications, anti-psychotics), psychiatry/psychology evaluation

COGNITIVE

- Cognitive enhancers (limited efficacy)
- Cognitive rehabilitation
- Compensatory strategies & enrichment

IMPAIRMENT IN ACTIVITIES OF DAILY LIVING

- Occupational therapy
- Home evaluation
- Devices to assist with ADLs (shower chair, shower bar, etc.)
- Long term care facilities





COMMON CHALLENGES IN THE CARE OF PERSONS WITH HD



OVERVIEW

- There are no magic formula involved in caring for a person with HD. You have the skills needed. You just need to learn a little about HD. Trust yourself!
- Don't touch the medications! Check with the HD care team or physician who prescribed them before making any changes.
- HD can be challenging. Remember, it's not the person with HD who is being deliberately difficult or unaware, it is the disease.
- Don't talk about the person with HD in front of them. Though they may not be able to communicate, they still understand everything you are saying.
- Talk TO the person with HD. Make eye contact, smile, and wait for them to respond. It may take a little while but they will!

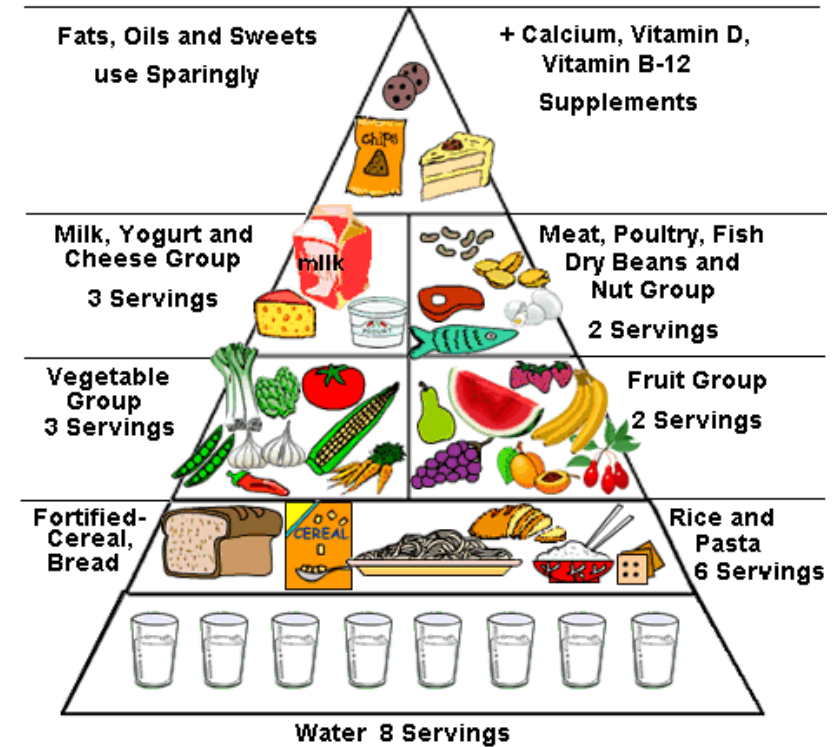
PROBLEM SOLVING

- What is the main problem?
- Gather information
 - ✓ When does it happen?
 - ✓ With whom?
- Review causes of behavior
- Give the person time
 - To speak
 - To process
 - To complete a task
- Set a realistic goal
- Be flexible and try several strategies
- Reassure all individuals involved
- As a patient is escalating, remember to de-escalate; do not feed into escalating behaviors
- ***Remember, it is Huntington's disease, not the person, that is causing the problem.***



NUTRITION AND SWALLOWING

- People with HD require significantly more calories than most residents in a LTC facility
 - A person with HD burns 5,000 calories/day
 - Sometimes, hunger or smells can lead to behavioral problems.
 - A PRN of pudding can pacify the hunger.
 - Double portions of meals may be required to help a person with HD maintain a healthy weight.
 - Many small meals throughout the day can provide needed calories.
 - Monitor patient's weight.
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- Some HD patients eat too fast, forget to chew or overstuff their mouths with food.
 - This increases the risk of choking.
 - Follow choking precautions in your facility.
 - Persons with HD should eat in a quiet area with no distractions.





A BIG BURST OF MOVEMENT

Persons with more advanced stages of HD often have difficulty controlling their voice and movements.

Common scenarios

- When getting up from a chair, the muscles and legs may use more force than needed to lift a person off the seat, giving the impression that they are leaping out of the chair.
- As you help them bathe, the person with HD may try to gently lift their arm to help you. Instead, they have a ‘big burst’ of arm movement. It may appear that you were hit, not helped.
- Think of using a bedside commode/toilet, grip bars, etc., to help with balance for patients when toileting. This can help avoid breakages from excessive movements.



PREVENTING FALLS

By the time the person with HD needs nursing home care, it is most likely that balance is impaired.

Pay close attention to walking and transfers to prevent falls.

If the person with HD has bedrails double check their safety.

Report falls to the resident's neurologist during visits.

REMEMBER

If balance is impaired, more falls are likely to occur while transferring into, onto and out of the bed, chairs and toilet.

REMEMBER

The goal isn't to prevent 100% of falls; focus on creating safe environments when falls are inevitable to prevent major injury.

MOTOR CONCERNS

For those persons with HD who have chorea, dystonia, or slowness, a special or padded wheelchair may be needed as walking abilities diminish.

You can also retrofit existing chairs or work with families to help provide specialty equipment.

- People with HD may have difficulty staying in a regular wheelchair because of their excess movement.
- PT or OT can help provide suggestions and documentation to access specialty equipment.

Uncontrolled movements may lead to the person with HD falling out of bed.

- The movement disorder can lead to a lack of sleep which can increase behaviors.



MANAGING BEHAVIOR

- Unawareness of symptoms
Is there a safety risk?
Remember, the person with HD isn't purposefully being difficult.
- Impulsivity
Is there a safety risk? If not, try to let it go.
Try to learn triggers; this is true for other symptoms as well
- Perseveration
Using distractions.
- Frustration, intolerance, anger
Avoid misinterpreting tone or body language.
No problem solving when patient enraged.
- Maintaining independence
When does ambulating or smoking become unsafe?
- Being right/Using reason
This tends to be a poor tactic, avoid using when managing unawareness, perseveration and frustration.





IMPULSIVITY

Difficulty in controlling impulses is caused by changes in the brain and not by the person being selfish or impatient.

Persons with HD cannot wait. Their brain is becoming cognitively rigid and the person can struggle with unknowns or fear of forgetting.

Persons with HD frequently have difficulty with tracking time.

STRATEGIES

- Respond immediately if you can.
- Utilize schedules to create routine.
- If you promise to help them in a minute, make sure you can really do it in a minute.
- Only make promises you can keep.

APATHY

It is hard for a person with HD to start or initiate an activity no matter how much they enjoy it.

Can be misinterpreted as depression.

The person with HD will continue to have the same interests as they used to.

STRATEGIES

- Find out from the person with HD and their family what they enjoyed before entering LTC
- Have staff encourage the person with HD to participate in activities
- Keep a structured routine as much as possible, and use visual reminders (calendar, to do list, etc.) to help remind a person to participate or complete scheduled activities.

Many LTC facilities have come up with creative ways to get the person with HD more active and involved.



PERSEVERATION

The person with HD may ask you the same question over and over.

Even though you take time to answer the question a few times, you may be asked again.

The person with HD understands what you are saying but has problems remembering or processing the answer, or cannot change the topic he/she is focused on.

STRATEGIES

- Please be patient.
- HD causes this behavior.
- Try to gently change this topic.
- Written schedules, reminders, dry erase boards can help alleviate constant questions



CHALLENGING BEHAVIORS

- Persons with HD may be irritable or aggressive. These behaviors are due to changes in the brain.
- While these behaviors can be based on actual problems or concerns, they are not an appropriate response.
- Utilizing reasoning or logic can exacerbate these behaviors or escalate irritability.
- Although sometimes the person with HD will acknowledge their reaction later, it is difficult for them to realize their overreaction in the moment.
- **Providing a safe environment for all is critical.**
- **Please work with your facility's psychiatrist and local HDSA clinic team to mitigate and manage these behaviors.**



Irritability and Aggression

A person with HD can have difficulty regulating their emotional response to stimuli.

Irritability can become sudden rage disproportionate to the stimulus.

Increased irritability can lead to aggressive episodes.

It can sometimes be difficult to redirect the behavior once it starts.


Aggression can often be managed through medication and environmental modifications.

Creating a structured environment and minimizing known triggers can help manage irritability and aggression

If the patient was being seen at an HD clinic, contact the clinic and HD care team when considering medication changes.

Did you know?

HD is excluded from psychotropic data reporting for LTC facilities



FRUSTRATION - CASE STUDY

A person with HD is located near the kitchen of the nursing home. Around 10:00 am each day she would start yelling loudly from her room and was an annoyance to staff and other residents. She had lost her ability to communicate.

STRATEGIES

The person with HD was reacting to the kitchen smells that increased her feeling of being hungry.

Once staff realized she was hungry, she received a mid-morning snack and the screaming stopped.

Alternatively, the person with HD could be moved to a room further away from the kitchen.



MAINTAINING INDEPENDENCE - CASE STUDY

A nursing home called the social worker about a young woman with HD who was “throwing herself out of her wheelchair”.

The social worker asked staff to explain what and where it happened. Staff never saw her do it but it happened in her room, in the bathroom and in the hallway.

Staff frequently found her sitting cross-legged and cross-armed on the floor controlling any extra movement.

STRATEGIES

- Facility identified this as a problem behavior.
- The person with HD was finding a position where she felt more in control of her body.
- Staff then worked at giving her opportunities out of her chair on a mat on the floor.
- The nursing home was able to stop writing incident reports about falls.

COGNITIVE DYSFUNCTION

- The cognitive disorder of HD affects a wide variety of mental functions.
 - It can take a person with HD more time to process information and respond.
 - A person with HD is sometimes unaware of his or her cognitive limitations.
- As the cognitive disorder progresses, the person with HD will lose the ability to organize his/her affairs, make decisions, and can be slow trying to do routine actions or learn new skills.
- The person with HD is generally aware of their surroundings, even if they are having problems responding to them.

Strategies

- Provide a consistent schedule. Make lists.
- Maintain a structured environment with limited distractions.
- Prompt each step of an activity with external cues.
- Reduce distraction.
- Give the person with HD extra time to answer.
- Ask close ended questions instead of open-ended ones, offer A or B choice options.
- Don't repeat while awaiting a response.



THE IMPORTANCE OF ROUTINE

Persons with HD have problems starting, continuing, finishing, planning and anticipating what is happening. Routine provides comfort and can reduce anxiety for the person with HD, which often impacts behaviors. Routine also applies to staff interacting with the person with HD, try to maintain consistency.

STRATEGIES

- Try to work out a routine of care for activities of daily living with the whole team.
- Set a schedule for bathing, dressing and eating.
- If doctor visits or other activities are not routine occurrences, tell the person with HD what will soon happen in order to avoid surprises.
- Limit changes in staffing.



THE IMPACT ON FAMILY

Although you're caring for the person with HD, you will interact with their family and it's important to remember that this is a family disease.

Take care with family members with whom you interact; this disease likely impacts multiple family members, some of whom may develop the disease.

STRATEGIES

- Try not to ask about risk status or genetic status. If a family member wants to talk about it, listen openly but try not to ask or pry.
- Practice empathy with family members; it is a family disease.



IMPORTANT TAKE AWAYS

1. HD is a complex disease with psychiatric, cognitive and motor symptoms
2. Persons with HD are not acting this way on purpose
3. Each person with HD is very different; do not base determinations about placement on past HD residents
4. Persons with HD cannot wait; you are not encouraging bad behavior by responding immediately, rather you are making both of your lives easier
5. HD families are your best source of ideas and help
6. Persons with HD deserve dignity; we should never give up on them or their disease
7. Good communication between the facility and the HD specialist will make everyone's life easier
8. Continue to facilitate ongoing HD care with their HD specialist(s)
9. No other facility has some special skills that you don't – each facility is capable of caring for persons with HD
10. There will be bad days...and they will pass



SUPPLEMENTAL MATERIALS

CASE STUDIES

STAFF INTERVENTIONS



CASE STUDY – MAINTAINING INDEPENDENCE

A man worked in the health care industry his whole life. When HD symptoms necessitated his move to the nursing home, he insisted on wearing scrubs every day and really believed he was an employee there. (Visitors sometimes made this assumption as well). Staff devised a system where he began delivering “notes” from one nurses’ station to the next throughout the day.

HD principle- Maintain independence
Encourage exercise
Sense of pride and importance
Increase socialization with staff and residents




CASE STUDY – MAINTAINING INDEPENDENCE

One family had similar HD traits of being arm-raisers, meaning that they would often have either one or both arms raised in the air when either sitting or walking. One family member was in a LTC facility where they were concerned about her frequent falls and her difficulty catching herself to prevent a fall. The staff was uncomfortable with her arm-raising symptom so they had instructed her to hold her hands behind her back to keep her arms down. They thought it would be uncomfortable or fatiguing to keep arms in the air all the time. So, even when walking around, she had been told to hold her arms behind her back.

HD principle- Maintain independence

Facility was uncomfortable with the excess movements of the woman though clearly a prominent HD symptom. The movements did not seem to bother the woman.

The patients' arm movements actually made her more stable and able to catch herself if she fell resulting in fewer falls.



CASE STUDY - IMPULSIVITY

A woman liked to go bed early (7-8 pm) in a very quiet and dark room. She was an avid pop drinker and went through multiple cans per day. Housecleaning staff person came into her room to empty her trash in the early evening making lots of noise as her pop cans were transferred to a larger bag on the hard tile floor. The patient woke, very agitated by the sudden noises and attacked the worker with a dull butter knife resulting in several staff persons needed to subdue her. She was sent to a psychiatric hospital and the nursing home refused to take her back.

HD principle- Impulsivity, Unawareness

HD patients can react/over react to stimulation like noise, light and activity.

Sudden changes can promote increased negative behaviors.

All staff should be present at in-services so that they are aware of these triggers.

What might have been a better strategy to handle this patient?



CASE STUDY - UNAWARENESS

A woman had a habit of slamming doors in two care facilities. In the first home the providers helped her decorate the door so it then became very special and the incidences of slamming the door decreased and then eventually stopped. When the patient moved to an adult family home the door slamming started up again. The caregiver worked positively with the patient to stop the behavior, and then decided to remove the door and simply replace it with a privacy curtain. The patient really liked the curtain!

Staff interventions- Unawareness

Instead of staff focusing on the specific behavior of door slamming, staff thought about ways to make the door special or make it disappear.



CASE STUDY – UNAWARENESS

A social worker shared her experience at an in-service where several of the staff were discussing how to treat one particularly irascible older woman. One caregiver said, “I don’t have much trouble with her. I just go in her room and talk to her about the day. I might touch her on the arm and describe what breakfast will be and what activity she might want to go to afterward.”

HD principle- Unawareness, Frustration

HD is a brain disease affecting a person’s ability to manage their frustration. The caregiver used distraction techniques to calm and mobilize the patient to participate in all activities.

This staff person seemed instinctively to know what the woman needed. This ‘born caregiver’ helped delivered her message to the rest of the staff.



CASE STUDY – PERSEVERATING

One man perseverated about a staff member at a facility and his desire for her to leave her family and marry him. He was very forceful and sometimes aggressive about his desires. He was also a lover of memories and his family helped him create a Memory Box with notes, letters and pictures from all of his family and friends. This box served as a good tool for NH staff to use for redirection. Whenever he got off track they suggested, "let's go find your memory box; I'd love to hear another story from there". He got so excited when someone wanted to hear about the memory box that he completely forgot about his desire for the married staff person.

HD principle- Perseverating

HD patients can get stuck on an issue and staff needs ways to “unstick” the patient and move on to other things.



STAFF INTERVENTIONS – CASE STUDY

A man in a LTC facility has a history of violent outbursts, though he does very well on certain psychiatric medications. Some state nursing home regulations require facilities to meet weekly or monthly to evaluate patients on psychiatric medications and adjust medications if behaviors are controlled. (Maybe these patients don't have bad behaviors because of the medications). But sometimes with a new nursing supervisor he was taken off his medications often resulting in him hitting a staff member. He was then sent to the hospital to readjust his medications and put back on the same dose of medication as he started.

Staff intervention-

Now they have a nursing supervisor who was willing to take the time to get the care plan carefully documented so that during their annual surveys they have clearly documented reasons to not lower or take him off medications.



STAFF INTERVENTIONS – CASE STUDY

A young man with HD was 6 ‘ 5”. He kept falling out of bed and crawling under the roommate’s bed or out into the hallway. He was depressed about his living situation but he also was extremely physically uncomfortable in a nursing home bed. He was evaluated and treated for depression but the getting/falling out of bed continued.

Staff intervention

Individual differences. He was a big man in a too small bed trying to find a proper position for sleep.

The nurse purchased a king size beanbag bed. This fit his large frame, was finally comfortable and the “behavior” stopped.



FINAL WORDS OF WISDOM

You are NOT alone! HDSA and our team of knowledgeable social workers, OT, PTs and dieticians are all here to help you. Call us!

ADD YOUR CONTACT INFORMATION HERE

THANK YOU FOR YOUR ATTENTION!

ACKNOWLEDGEMENTS

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