Partners in FTD Care

a quarterly newsletter for healthcare professionals

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FTD Symptom or Pain - How Can You Tell?

The unusual behavior of someone with frontotemporal degeneration (FTD) is not always what it seems. Because people with FTD are often unable to express their needs verbally or accurately, treatable medical conditions and pain are easily missed causing unnecessary distress to the patient and complicating care management.

First back pain, now FTD

Joan Brown is a disabled 63-year-old college professor who suffers from moderate-to-advanced frontotemporal degeneration (FTD) with significant problems with behavior and language.

Dr. Brown married and had twins in her late 30s. Lean and active, Dr. Brown ran track and played tennis during high school and college. She developed back pain while carrying her twins but began to run marathons and continued until she developed FTD.

At age 53, she completed menopause and became emotional. Her behavior became more impulsive and she began to "pull away" from her family. Students complained that her behavior towards them was inappropriate. This was attributed to "the change" and estrogen replacement was tried without success. Routine bone density scans revealed osteoporosis. Over the next three years, x-rays done for pain revealed degenerative joint disease and degenerative disk disease in her spine. She sought chiropractic care and acupuncture routinely for pain management.

Dr. Brown's initial symptoms of FTD were identified at age 56 when she developed agrammatic /non-fluent primary progressive aphasia. She was able to understand written and spoken language but lost the ability to produce words. Early in the disease, she experienced great difficulty lecturing. The first few years after diagnosis, she was able to continue teaching via computer as she was able to type her lectures. At age 58, she could no longer communicate verbally with colleagues and retired from teaching.

Increasing FTD symptoms

At age 61, Dr. Brown became mute. She no longer understood what people were saying to her. She no longer complained of pain, and when asked if she had pain, she shook her head "no." Believing the pain had resolved the family stopped the pain treatments.

Her behavior was impulsive and she was unable to perform even simple household chores. Her grooming had suffered, and she had frequent urinary and fecal incontinence when in a mall or social situation but refused to wear incontinence garments. She spent her days roaming her neighborhood, occasionally going too far, necessitating the rescue squad for assistance locating her. Her husband confined her to their home where she roamed 12 to 14 hours per day in a precise pattern. She resisted her husband's help with foot and toenail

Partners in FTD Care *is an education initiative of the Association for Frontotemporal Degeneration that brings together health professionals, experts and families to promote understanding of FTD and to develop best practices in community care.*

Did You Know?

Fully 70% of people with FTD are diagnosed under the age of 65. Estimates of the prevalence of FTD show its frequency in the population approaches that of young onset Alzheimer's disease.

Approximately 2-5% young onset Alzheimer's is caused by genetic mutations that run in families. In FTD, 10-20% of cases are caused by familial genetic mutations. The generous participation of these families in research is driving scientific discovery forward.

Medical centers are starting to recruit participants for the first clinical drug trial of a potentially disease modifying treatment for behavioral variant FTD (bvFTD).

Contact Us!

For more information on FTD and advances in research and care contact AFTD's HelpLine (info@theaftd.org or 877-507-7222) and register with AFTD--www.theaftd.org.

care. When she stopped sleeping at night at age 62, her family decided to place her in a memory care facility.

On admission to memory care, Dr. Brown continued her roaming with an increasing sense of urgency as days progressed. She began to yell spontaneously and was resistant to care. The staff contacted her doctor for something to calm her down. Ativan was prescribed, but it worsened the agitation and was stopped. The facility asked for an antipsychotic. Both risperdone and quetiapine were tried with minimal success. Citalopram, an antidepressant was also tried and failed to help.

Three weeks ago, Dr. Brown fell while trying to climb out of a window. She was sent to the ER where staff and family questioned her repeatedly about pain. She always responded by shaking her head "no," and was returned to the facility. Staff reported she was becoming aggressive and resistant to care. Whenever approached by staff towards her right she would strike out. It became impossible to dress her. Her vocalizations increased to screams, especially at night. When no one was near her, she would retreat to bed and moan occasionally.

A sharp CNA's observation

Staff noted her grimacing during mealtime. Her primary nursing assistant commented that she felt Dr. Brown might be in pain, although her idea was dismissed as she continued to deny it. The staff considered a referral for an inpatient acute care psychiatric admission to manage "her aggressive problem behaviors," when her CNA spoke up again: "I think Dr. Brown is in pain because she won't let me touch her right side." An X-ray of her right arm and shoulder revealed a fracture of her clavicle (collarbone). An orthopedic surgeon ordered a right arm splint to be worn at all times and prescribed one gram of acetaminophen, three times a day. Staff was much more careful not to move or jostle Dr. Brown's right arm and shoulder when providing care. They were careful to approach her from the left and not use her right shoulder when helping her to move. Gentle heat was provided for comfort.

Dr. Brown stopped vocalizing and once again allowed her CNA to bathe and dress her. The staff began using the PAINAD Scale to evaluate for the presence and intensity of pain. With Dr. Brown's history of chronic back and joint pain, the providers and care staff decided to treat the pain by preventing it. The acetaminophen was continued indefinitely and the staff monitored carefully for "breakthrough" episodes of pain. A physical therapy consultation was ordered to assist with maintaining joint mobility and for use of modalities such as massage or electric stimulation to minimize pain.

Questions about the case:

1. **How did Dr. Brown non-verbally express pain?**

Dr. Brown's non-verbal expressions of pain included: roaming,

TROUBLES & TIPS

Q: We see people with FTD who have many unusual behaviors that are not common in other types of dementia. The behaviors vary in their impact on functioning day to day and change over time. How can we tell if someone with FTD is experiencing pain?

A: Significant changes in personality, behavior and language are hallmarks of the FTD spectrum disorders. These characteristic symptoms don't occur in predictable stages, but can increase or decrease as the disease progresses.

Automatically assuming a new or changing behavior is due to FTD may cause families and providers to miss treatable medical conditions masked by the underlying neurological disorder. Evaluating pain through observation of new or unusual behaviors, using non-verbal assessments, and receiving and integrating feedback from the family and caregivers are keys to identifying and managing pain.

It is often easier to recognize non-verbal communication and behavior change in people with Alzheimer's disease (AD). When someone with AD has a sore foot or muscle pain, they stop walking. Someone with FTD who has compulsive behavior symptoms will typically keep walking even if their foot hurts. People with AD typically retain a range of facial expression. FTD patients may not recognize emotion from faces and may not show emotion through facial expression.

The two main evaluations used to assess pain for a non-verbal person require numerical pain scales (PAINAD) or rating facial expression (Wong–Baker FACES). While PAINAD has been validated in the AD population, neither has been validated for use in FTD and may not be reliable.

Even general or vague reports from family or direct caregivers that something is different or not right should prompt a nursing evaluation. This is particularly important after an incident, such as a fall. In some cases, the medical team may treat with acetaminophen and observe for reduction in behavior to assess for unidentified pain.

Careful observation and common sense effort will improve the person's quality of life and reduce disruptive behavior.

resisting care and yelling spontaneously, all with increasing urgency over time. She was more sensitive to touch on her right side, grimacing, striking out when approached from that side, moaning, retreating to bed, screaming and increased agitation.

2. Why was Dr. Brown's pain initially not treated?

Dr. Brown was an unreliable source of information for pain evaluation. When asked if she had pain, she answered "no." Individuals with FTD and other dementias with loss of verbal skills may be unable to understand and respond accurately to questions. They may automatically respond "no" to questions they do not comprehend (as in Dr. Brown's case). Due to Dr. Brown's negative responses, the caregivers' observations of her pain were dismissed.

While behavior changes are recognized as a hallmark of FTD, many of the behaviors seen in a person with moderate to advanced FTD (i.e., refusing dressing or personal care, compulsive walking or altered sleep patterns) could also have a medical cause. Behaviors that might trigger a nursing assessment in someone with Alzheimer's disease are often attributed to the FTD itself. Dr. Brown's changes in behavior were attributed to increased agitation and depression. Several medications, including an antipsychotic, were tried with no success.

3. What effective pain evaluations and treatments were provided for Dr Brown?

The CNA's observation of Dr. Brown's reactions (grimaces, sensitivity to touch on the right side) and assertive communication of a suspicion of pain prompted a medical evaluation. Dr. Brown received an X-ray, orthopedic consult and treatment of the fracture that included a splint, routine acetaminophen and gentle heat. Facility staff used specific care techniques, i.e., approach from the left, avoid stress on the right shoulder and gentle movement. They then took a preventive approach to pain using the PAINAD Scale (see Resources section) for ongoing observation of pain, routine acetaminophen and physical therapy techniques.

4. What history of physical conditions contributed to Dr. Brown's pain?

Dr. Brown developed back pain while carrying her twins. She was a marathon runner. She has a history of osteoporosis, degenerative joint disease, and degenerative disk disease. Recently she had a fall. Her compulsive walking and poor foot care may have also caused pain for some time prior and exacerbated care management challenges.

5. How could that information have informed more proactive care?

Her family and caregivers did not understand the importance of Dr. Brown's history of painful physical conditions once she was diagnosed with FTD. Such awareness may have prompted them to question her denial and look more closely at behavior changes. Had the caregiving staff noted her history of physical symptoms, (chronic back and joint pain) and prior treatment (acupuncture and chiropractic care) they may have recognized the presence of untreated pain earlier. Use of AFTD's Partners in FTD Care Daily Snapshot Tool is one way to capture critical history from the family prior to placement. Dr. Brown resisted personal care at home, was not sleeping, roamed and was unable to communicate. This information would have provided a baseline for evaluating and managing Dr. Brown's pain after transition to the facility.

RESOURCES TO CHECK OUT

Tools for Assessing Pain

Two pain evaluations used for a non-verbal person include:

The PAINAD (Pain Assessment In Advanced Dementia) Scale is a numerical scale that includes non-verbal indicators of pain, such as body language and facial expressions. It should be completed while the person is at rest and during movement.

http://www.healthcare.uiowa.edu/igec/tools/pain/PAINAD.pdf

The Wong-Baker FACES Pain Rating Scale is a scale that includes facial expressions indicating pain from "No Hurt" through "Hurts Worse."

http://www.wongbakerfaces.org

AFTD's Daily Care Snapshot Tool (Family/Caregiver Informational Tool) communicates information regarding the individual's past and current physical conditions, treatments, care needs, behaviors, etc., to community care providers.

http://www.theaftd.org/wp-content/uploads/2011/09/Packet-Daily-care-snapshot.pdf

Evaluating and Managing Pain

Evaluating pain in people with frontotemporal degeneration (FTD) can be challenging. Behavioral changes are often attributed to the disease rather than triggering an evaluation for pain or discomfort. When you understand FTD symptoms and adopt an observant approach, multi-disciplinary interventions can make a real difference in compassionate care.

Tips for identifying and managing pain include:

- Anticipate pain from concomitant conditions such as arthritis, back pain, peripheral neuropathy and other medical conditions. Treat.
- Use AFTD's Daily Care Snapshot Tool (http://www.theaftd.org/wp-content/uploads/2011/09/Packet-Daily-care-snapshot.pdf) to communicate a person's medical issues and past treatment to care providers.
- Don't discount the ability of the person with FTD to give an appropriate response to questions, but confirm with additional evaluation.
- Avoid "Yes/No" questions when asking about pain. People with FTD may answer indiscriminately; the first response is usually "No."
- Ask specific questions, e.g., "Where is your stomach?" or "Press where it hurts." Include gestures to provide non-verbal cues, e.g., point to stomach, etc.
- Automatically check for range of motion. For example, during personal care such as dressing, routinely check feet and shoes of people who roam or pace extensively.
- Non-verbal pain scales that use numbers or faces may be helpful, but don't stop after a single assessment. People with FTD may not recognize emotion from faces use in the facial pain scale.
- Don't discount general or vague reports from family or care providers that something is different or not right. Ask them to look back in time; incremental changes may not be noticed day to day.
- Treat with routine acetaminophen to assess vague reports of something being different; if the behavior subsides, it may confirm the presence of pain.
- Non-verbal behavior is more difficult to read in FTD than Alzheimer's disease (AD). In AD, behavior is generally consistent. Someone with a sore foot may stop walking which triggers evaluation. A person with FTD and compulsive behavior may keep walking even if it hurts.
- Increased chewing behavior or decreased appetite and eating may indicate pain in the teeth/gums and should be evaluated.
- Watch for resistance to dressing or personal care which may indicate pain; evaluate. If reaction is in the shoulder, switch from a pullover to shirt with buttons.
- Grabbing behavior (a person's wrist, arm, etc.) may be common in FTD as related to a frontal lobe reflex. If it intensifies, it may be a reaction to pain.
- Someone scratching at his or her crotch may have pain or itch in that area rather than disinhibited behavior. Skin evaluation (especially of the peri area) should be done each bath day.
- Develop a pain management plan and communicate it to all staff and the family for input and to provide consistent care.
- Include PT, OT and speech therapies in pain evaluation and management, i.e., exercises, heat/cold, massage, positioning, transfer techniques, mobility beds/devices and adaptive clothing/devices.
- Provide recreational interventions, such as soothing music, pet visits, aromatherapy and massages.
- Use common sense and careful observation on a continuing basis.

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an education initiative from AFTD

Expected FTD Behavior or a Superimposed Medical Condition?

Significant changes in personality, behavior and language are hallmarks of the frontotemporal degeneration (FTD) disorders. These characteristic symptoms do not occur in predictable stages and can increase or decrease as the disease progresses. Assuming a new or changing behavior is due to FTD may cause families and providers to miss treatable medical conditions masked by the underlying neurological disorder.

FTD behavior or symptom		Possible medical considerations
Apathy, social withdrawal, reduced initiative	May not eat or drink properly if not monitored	Weight loss, dehydration, electrolyte disturbance
	Decreased personal hygiene	Altered skin condition, rashes, infections; urinary tract infection; tooth decay
	Inadequate physical activity	Constipation; weight loss, generalized deconditioning, failure to thrive; blood clots due to circulation problems
Emotional blunting, loss of empathy	Hallmark of FTD, also in depression	Possible manifestation of pain
Disinhibited behaviors	Compulsive eating , especially carbohydrates	Weight gain and increase in associated health risks; tooth decay
	Compulsive drinking of water	Electrolyte disturbance presenting as increased confusion
	Compulsive drinking of caffeinated beverages	Agitation; elevated blood pressure and pulse; electrolyte disturbance; urinary incontinence
	Stuffing food into mouth	Death from choking; aspiration pneumonia, lung infections
	Hands in pants; scratching at crotch	Skin irritation, rash; urinary tract or other infection , vaginal yeast infection, prolapse
	Disruptive vocalizations, yelling	Generalized pain
	Disrobing (woman)	Hot flashes, hormone changes from menopause
Repetitive or stereotyped behaviors	Constant walking or roaming	Foot blisters, infections of feet; muscle pain; trauma from tripping or falling; danger of elopement
	Picking or biting fingers	Hand infections, tearing of nail beds and bleeding
	Chewing behavior	Tooth or gum pain
Impaired language fluency	Decreased verbal output expected with progression of PPA	Possible sign of mouth or teeth pain
Comprehension impairment	Inability to understand word meaning and concepts, recognize faces and objects	Vision changes requiring eye exam, updated glasses
	Unlikely to understand common scales used to evaluate pain	Close behavioral observation, non-verbal assessments
	May not recognize or conceptualize painful sensations	Monitor cooking activity, exposure to fire, heat, cold other hazards; evaluate after exposure
Movement or motor skills impairment	Abnormal limb posture(ie: CBD)	May indicate painful contractures, frozen shoulder from lack of use, fracture
	Unsteady gait and balance predisposes to falls	May indicate blisters, poorly fitting shoes
	Sudden cessation of walking and mobility	Assess for pain
Changes in mood	Agitation	Assess for pain; overstimulation or changes in environment; fatigue; excess caffeine
	Sadness or depression	Clinical depression (especially in PPA); may be sign of pain
Emotional incontinence common in PSP	Forced laughing or crying out of context	May indicate bodily sensation that needs investigation