ALS® ASSOCIATION
St. Louis Regional Chapter

PROVIDING HEALTH, HELP, AND HOPE

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Amyotrophic Lateral Sclerosis

Medical description

- Amyotrophic Lateral Sclerosis (ALS) is a fatal, neurodegenerative disease.
- Often known as Lou Gehrig's disease or motor neuron disease, it is a progressive disorder that occurs when motor nerve cells in the nervous system cease functioning and die.
- Muscle control becomes completely lost, resulting in paralysis.
- Death normally occurs due to secondary infection or insufficient ability to expel carbon dioxide.

Lay person's description

- Motor neurons carry the message from our brain and spinal cord to our muscles in order for us to perform any movement.
- In ALS, the motor neurons die and the message is not given to the muscles for movement.
- You may want to comb your hair or take a step, but your muscles do not get the message and movement thus becomes difficult and eventually not possible.
- Most of the time a person's mental capabilities are not affected.
- ALS does not affect the senses.

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Types and Forms

Types of onset

Limb onset

Begins in legs or arms and "spreads" thus affecting all limbs before moving to the bulbar region.

Bulbar onset

Begins in the bulbar region, (located in the brain stem at the base of the head), with speaking and swallowing difficulties. ALS will then move to the limbs.

Forms

- **Sporadic**
  - Most common form.
  - 90% of all cases do not have a genetic predisposition.
- **Familial**
  - Up to ten percent of ALS cases are familial.
  - One form linked to chromosome 21 – SOD1 gene.

Frontotemporal Lobe Dementia (a change in personality and in mental processes), can occur together with ALS.

Symptoms of ALS

- Fatigue
- Respiratory concerns
- Muscle weakness, stiffness, atrophy
- Muscle spasms
- Poor fine-motor coordination
- Swallowing difficulties, chewing fatigue
- Pseudobulbar Palsy
- Tongue weakness
- Weight loss
- Speaking difficulties
- Change in voice quality
- Sialorrhea (drooling)
- Mobility difficulties
- Constipation
- Depression

COMMUNICATION AND SPEECH: PROGRESSION OF LOSS

When your client begins to have trouble with speech and communication you may notice:

- Hoarse or raspy voice
- Speaking through the nose (nasal speech)
- Difficulty with long conversations

As speech and communication issues worsen, you may notice:

- Talking becomes exhausting
- Difficulty with long sentences
- Difficulty projecting voice
- Slurred speech, or difficulty forming words

When speech and communication issues advance, your client will:

- Lose the ability to speak all together
- Rely on assistive technology to express themselves

COMMUNICATION AND SPEECH: CARING FOR THE CLIENT

- Give the person your full attention-concentrate on the face while talking
- Use yes, no, or maybe questions
- Never rush the conversation
- If you don't understand something, repeat the portion you don't understand and ask for clarification
- Ask the speaker to spell or change words that aren't clear
- Ask the speaker to indicate when the topic is changing so you'll know the context

- Repeat what you understood so the client can simply fill in missing words
- Talk in a relatively quiet space-turn off any external noises (TV, music, etc.)
- Encourage the client to take a deep breath- this provides "fuel" or power to the words
- Establish a limit on how many times you will ask the client to speak their thoughts before you use another method of communication

COMMUNICATION AND SPEECH

LOW TECH COMMUNICATION TOOLS

- Use head nods, eye blinks, gestures or sign language
- Encourage communication through writing: dry erase board, note pad, or tablet.
- Encourage the client to write only key words to help conserve energy
- Have them point to letters or pictures using finger or laser pointer

HIGH TECH COMMUNICATION TOOLS

- Computers with voice synthesizers
- Communication applications (word completion, word prediction)
- Remote control units for environment
- Eye-gaze Response Interface Computer Aid

Always remember.. The client's communication needs will change overtime.

COMMUNICATION AND SPEECH: IMPACT

- Losing the ability to communicate causes the feeling of isolation from family, friends, and community.
  - If a client has also lost the ability to move, thinking and expressing thoughts may be the only bit of independence left.
- Losing the sound of a loved one's voice can cause sadness and grief for family and friends. The burden of working to communicate without speech often causes frustration for the client, the family, and caregivers.
- Finding ways to assist with communicating their thoughts and needs will help the client interact and allow them to direct their care.
  - Being able to do this will hopefully provide them with the feeling of dignity and independence

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MOBILITY:
SIGNS AND SYMPTOMS

When your client begins to experience issues with mobility, you may notice:

- Spasticity, muscle cramping
- Muscle twitching (also known as fasciculation)
- Uncoordinated movements (can lead to falls and ability to stop falls)
- Fatigue in performing simple tasks, such as ADLs

As mobility issues worsen, you may notice:

- Some muscles are paralyzed
- Fasciculation continues
- Clients may have lost the ability to perform ADLs

As mobility issues advance, you may notice:

- Client is paralyzed
- Can still feel muscle cramping, pain, and discomfort

MOBILITY:
FINE AND GROSS MOTOR SKILLS

FINE MOTOR FUNCTION

The coordination of muscles, bones, and nerves to produce small, precise movements. Examples:

- Buttoning a shirt
- Zipping a zipper
- Picking up a fork
- Writing
- Typing
- Dialing a phone

GROSS MOTOR FUNCTION

The opposite on fine motor movements are larger and less precise. Examples:

- Walking
- Climbing stairs
- Waving of the arms
- Reaching
- Maintaining balance
- Turning in bed

MOBILITY:
CARING FOR THE CLIENT

What can you do to help with mobility?

- As mobility issues increase, maintaining a safe environment and preventing injury are important aspects of the HHA's role.
  - Monitor the client for increased muscle weakness and communicate concerns with the family
- As ALS progresses, the client will require assistance with Range of Motion (ROM) exercise and transferring to and from their bed, chair, toilet, shower, or into the car.
- Always remember… despite the loss of mobility, the client still feels sensation such as pain and irritation
- Shrinking and wasting away of muscles causes ALS clients to lose the ability to move, and this occurs at different rates for all clients.

MOBILITY:
TIPS FOR SAFE TRANSFERS

- Encourage the client to participate in their care to the best of their ability and never force movement
- Always make sure you know how to use any equipment properly
- Maintain proper body mechanic when transferring or repositioning the client
- Don’t rush the client
- If transfers are difficult or involved, break down the transfer into smaller parts
- Clearly verbalize each step to the clients as you transfer them
- Maintain clear pathways and remove all safety obstacles
- Keep all assistive devices within reach of the clients
- Lock the breaks on wheelchairs and beds before moving the client and confirm all connection before using a lift

Always follow the care plan

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SWALLOWING AND NUTRITION: SIGNS AND SYMPTOMS

PHASES OF NORMAL SWALLOWING

- Tongue works to keep food and fluids in the mouth
- Food is chewed and mixed with saliva
- Food is moved from mouth to throat
- Some muscles open to allow food into the esophagus while other close to keep food out of the windpipe
- Food enters the esophagus
- Muscles in esophagus push food down to stomach

SIGNS YOUR CLIENT IS HAVING TROUBLE

- Excessive drooling
- Complaints of difficulty or discomfort swallowing
- Pocketing or storing food in the mouth while eating
- Coughing frequently when drinking or eating meals (aspiration)
- Changes in amount of types of food the client is eating
- Regurgitation of food into nose, throat, or mouth
- Difficulty/inability to swallow
- Complaints of pain or discomfort when swallowing

SWALLOWING AND NUTRITION

TIPS FOR ASSISTING WITH ORAL FEEDINGS:

•Provide high calorie foods in small amounts

•Use thickened liquids to alter the consistency, making it easier to swallow

•Incorporate foods high in fiber

•Provide high protein enriched supplements

•Encourage small bites

•Provide plenty of time for meals

•Add snack throughout the day

•Support use of adaptive utensils

•Provide soft foods like pudding or applesauce

CARING FOR THE CLIENT:

The role of the professional caregiver is to maintain safety and support

•Communicate changes to the family

•Support and follow dietary changes when providing meals

•Prevent choking and aspiration by following suggested feeding techniques

•Encourage fluid intake in order to prevent thickening of the saliva

•Provide a balanced diet with adequate protein

ELIMINATION:
SIGNS AND SYMPTOMS

When your clients begins to have trouble eliminating independently you may notice:

- Decreased liquid intake
- Difficulty getting to the bathroom on time
- Foul odor to urine- early sign of UTI

As issues worsen you may notice:

- Difficulty passing urine or stool
- Constipation
- Darker/amber colored urine
- Weight loss

When issues become dangerous, your client may have:

- Impacted bowel movements
- Bowel leakage
- Dehydration and malnutrition
- Fever
- Pain

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ELIMINATION:
CARING FOR THE CLIENT

DIET

- Encourage increased daily fluid intake
- Be familiar with products used to maintain healthy bacteria in the gut
- Be aware of how changing diets can affect the consistency and frequency of bowel movements, such as severe diarrhea or constipation.
- Provide nutrient dense foods (salmon, shellfish, eggs, kale) that are high calorie, high energy, and low volume

ELIMINATION

- Watch for constipation, diarrhea, and impaction
- Be organized and plan to have all supplies and materials on hand
- Keep bedside commode or urinal nearby
- Use strict hygiene practices around catheters
- Keep skin clean and dry to prevent pressure ulcers from developing.
- Know the client's elimination habits
- Watch for signs of UTI: urgency, painful urination, fever.
- Respect the client's privacy and dignity. Handle incontinent problems quickly and respectfully.

Red Flags:

- Change in form, odor, color, and consistency of stool or urine
- Abdominal, mid-back pain, or discomfort
- Confusion
- Nausea, vomiting, or diarrhea

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PAIN AND COMFORT MANAGEMENT: CLIENT COMFORT

- **Keeping the client comfortable is very important. Imagine sitting or lying in one position for an extended period of time and unable to change positions to relieve pressure.**
- **Pain and comfort may be reduced through frequent repositioning and range of motion exercises (ROM)**
- **Tuning in and communicating with your client will help you become more aware of the subtle cues.**
- **As the disease progresses and muscle function is lost, pain increases**
- **Emotional pain can worsen physical pain, and vice versa**
- **Since pain is not a direct consequence of ALS and is not always reported by the client, it is often improperly managed.**
  - This causes needless suffering
- **Poorly managed emotional and physical pain can have a devastating effect on the emotional and social wellbeing of clients with ALS**

PAIN AND COMFORT MANAGEMENT: COMMUNICATION

Remember that your client may not be able to physically express their pain because the muscles in their face are too weak. Make sure to frequently ask *"Are you having any pain?"* and ask the client to rate their pain using the 1-10 pain scale

Pain Scale
Using the 0-10 scale, rate the change in severity in an effort to monitor levels of pain; 0=no pain, 10=most severe

Descriptions of Pain
Achy – acute – agonizing – burning – dull – excruciating – gnawing – gripping – irritated – raging – raw – severe – sharp – sore – stabbing – stiff – tender – tight

RESPIRATORY:
SIGNS AND SYMPTOMS

Beginning of respiratory problems, you may notice:

- Loss of energy
- Weakened or soft voice
- Weak, quiet cough
- Frequent clearing of throat
- Difficulty projecting voice
- Trouble sleeping, walking

As problems worsen, you may notice:

•Frequent clearing of throat

•Orthopnea (the need to sleep upright)

•Excessive Fatigue

When problems escalate beyond control, clients:

- Lose the ability to breathe on their own
- Have respiratory failure without mechanical support

RESPIRATORY:
CARING FOR THE CLIENT

How can you help a client with breathing difficulties?

- Avoid exposure of any type of cold, virus, or flu- keep client away from people who show such signs
- Avoid dust and fumes including perfumes, aerosol sprays, powders
- Position client with pillows to maintain alignment of the spine
- Optimize breathing with elevating head and have client sit upright before, during, and after meals
- Avoid overeating (can worsen breathing)- encourage smaller meals, and more frequent meals and snacks throughout the day
- Ensure proper fluid intake to help keep secretions thin and easier to cough up or suction out

RESPIRATORY:
ASSISTED VENTILATOR OPTIONS

Cough Assist:- a small machine used to clear airways and remove secretions from lungs. It helps reduce occurrence of respiratory infection and is a safe, noninvasive alternative to suctioning

Ventilator- does all the work of the respiratory muscles. A breathing tube is surgically placed through a hole in front of the neck and into the windpipe.

BiPAP- a small machine used to assist with airflow into and out of the lungs, a small mask is applied to the client’s nose or mouth. Most begin using the BiPAP at night and progress on and off during the day

PSYCHOSOCIAL IMPACTS

Grieving the ALS diagnosis and the losses that it brings is normal. Normal and natural emotion responses for client and family caregivers throughout the disease processes may include:

- **Anxiety**
- **Fear of the unknown**
- **Loneliness**
- **Fear of abandonment by family and friends**
- **Fear of suffering and pain**
- **Anger and hostility**
- **Guilt and shame**
- **Denial or shock**
- **Depression and sadness/suicidal thoughts**
- **Remorse**
- **Withdrawal**
- **Crying**
- **Hopelessness**

PSYCHOSOCIAL IMPACTS:
CLIENT AND FAMILY CAREGIVER LOSS

Loss	Client	Family Caregiver
Independence	Inability to perform self-care and activities (Driving, cooking, cleaning)	Missing out on social activities; missing work
Control	Inability to stop changes happening to their body; loss of control within family	Inability to control physical and emotional losses; inability to change outcome of ALS
Employment/finances	Must depend on other financially	Financial strain due to loss of income; loss of health insurance
Dreams/hopes for future	Future plans dismissed; sense of missing out on important life events	Future plans interrupted; emotional having to think about future without loved one

PSYCHOSOCIAL IMPACTS CONTINUED

Loss	Client	Family Caregiver
Self-identity and self-esteem	No longer the person they used to be; unsure of who they will become as ALS progresses	Change in dynamics of marital relationships; loss of intimacy; not connecting in familiar ways
Life purpose and meaning	“Why am I here?” “Why did this happen to me?”	“What did I do to deserve this?” “Why is this happening to me?”
Social life and relationships	Difficulty relating to loved ones in the same way; dependence on others; missing out on social activities	Missing out on social activities due to caregiver responsibilities’ no longer relate to family and friends
Sexual intimacy	Fatigue, muscle weakness, incontinence, negative self-image	Sees shift in role from lover to caregiver; no longer feeling connected

CONCLUSION:
CARING FOR THE CLIENT

As a professional caregiver, you can:

- Allow and encourage the client and family to communicate their thoughts and feelings
- Be a good listener; listen first, develop an understanding, then respond
- Encourage the client to be hopeful
- Respect privacy at all times
- Communicate changes to the family
- Respect any religious customs
- Encourage the family caregiver(s) to utilize the time the loved one is with a trusted caregiver to experience self-care or social engagement outside the care environment

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It's Not Just Lou's Disease

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