

WELCOME !

December 6, 2021



The ALS Association

National Office-Care Services

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ALS Multidisciplinary Care

Guest Speaker:

Lauren Tabor-Gray, PhD, SLP-CCC

Co-Director, Phil Smith ALS Clinic

and Clinical Research Center

Clinical Scientist, Department of Neurology

Holy Cross Health

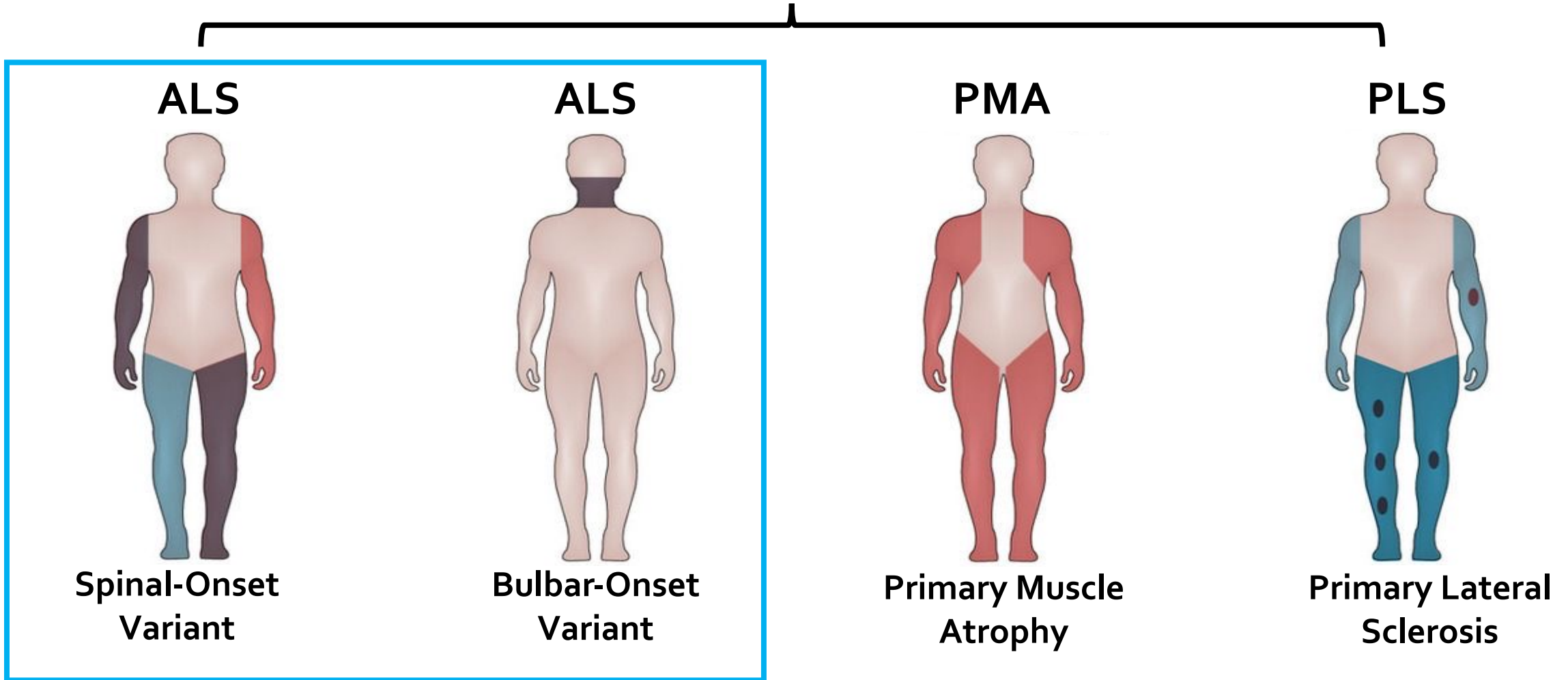
Optimizing ALS Care:

the Multidisciplinary Clinic

Lauren Tabor Gray, PhD
Co-Director, Phil Smith ALS Clinic
Clinical Scientist, Dept. of Neurology
Holy Cross Health

12/6/2021

Motor Neuron Disease



Amyotrophic Lateral Sclerosis

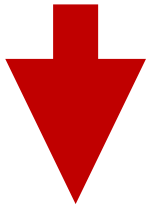
- A: No ; myo: Muscle; trophic: nourishment
 - *No Muscle Nourishment*
- Lateral: denotes the area of nerve cells affected (both upper and lower motor neurons)
- Sclerosis: or *hardening* that occurs as the motor neurons degenerate

ALS Pathology

UMN

Cortical Motor Neurons
Corticospinal/Bulbar Tracts

Supranuclear Symptoms



Spasticity

- Muscle Stiffness
- Muscle Slowness
- Hyperreflexia
- Decreased speed



LMN

Brainstem
Cranial Nerve Nuclei

Bulbar Palsy



Atrophy

- Flaccid Paresis
- Decreased Strength
- Decreased Force
- Fasciculations

What does *evidence-based* care
look like for pALS?



- Patients as Partners
- Friends and Family
- Caregivers
- Primary Care Doctors
- Hospital and Emergency Room
- Palliative and Hospice
- Home and Outpatient therapists
- Home Health agencies

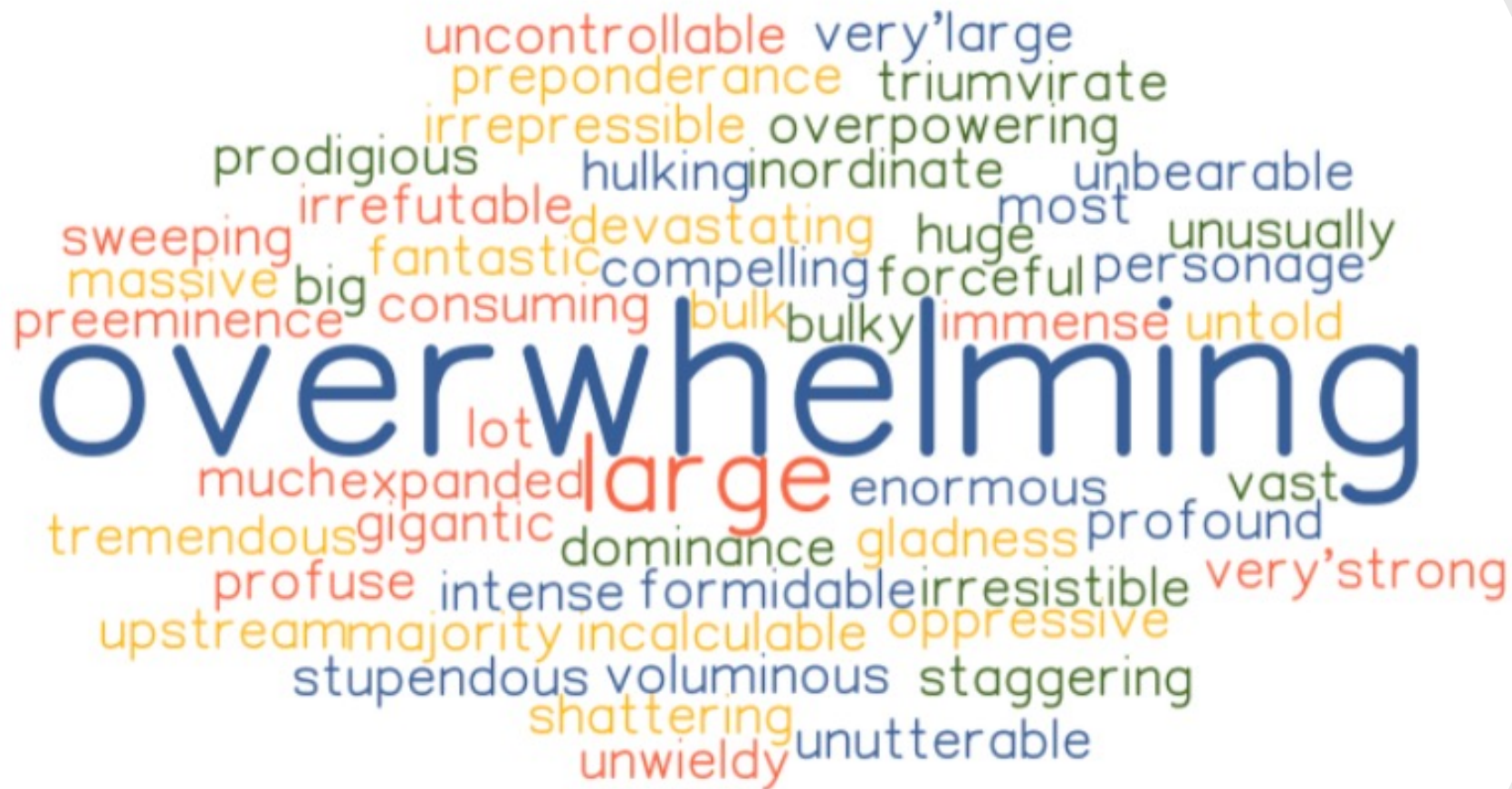
the Mission: Comprehensive Care

- Multidisciplinary clinics
- Multidisciplinary Telehealth appointments
- Mobilize Community Resources: Partnership with MGH, ALSA, MDA, Always for ALS
- Advocacy: Clinical Research, ALSA, ALS TDI, Legislation, Patient Ambassadors (CURLI)

The Team

- Neurology
- Nurse Practitioner
- Nurse/Clinic Coordinator
- Speech Language Pathologist
- Occupational Therapy
- Physical Therapy
- Dietitian
- Social Work/ALSA Liaison
- Assistive Technology Specialist
- Neuropsychologist
- Palliative Care Team
- Research

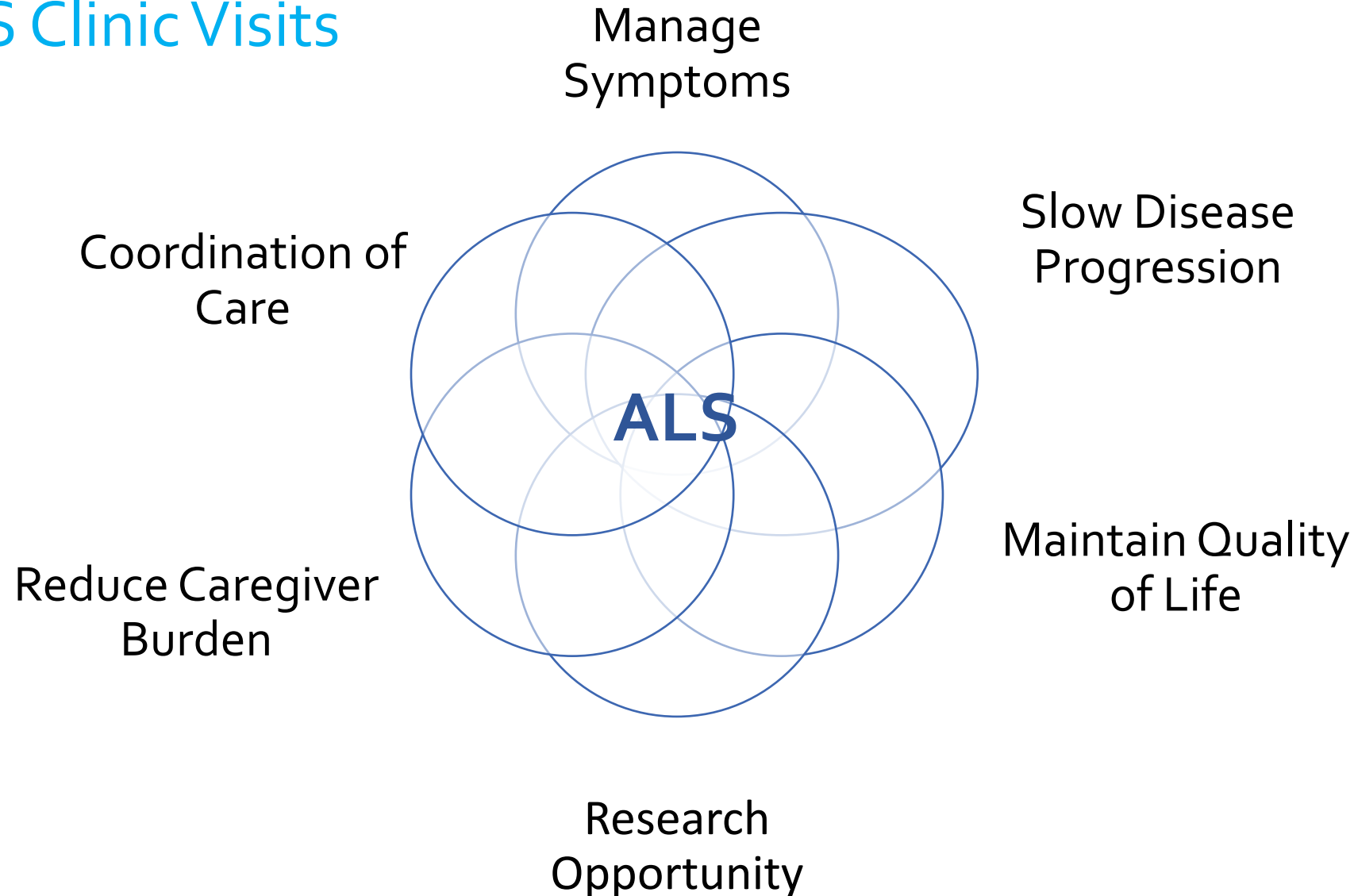




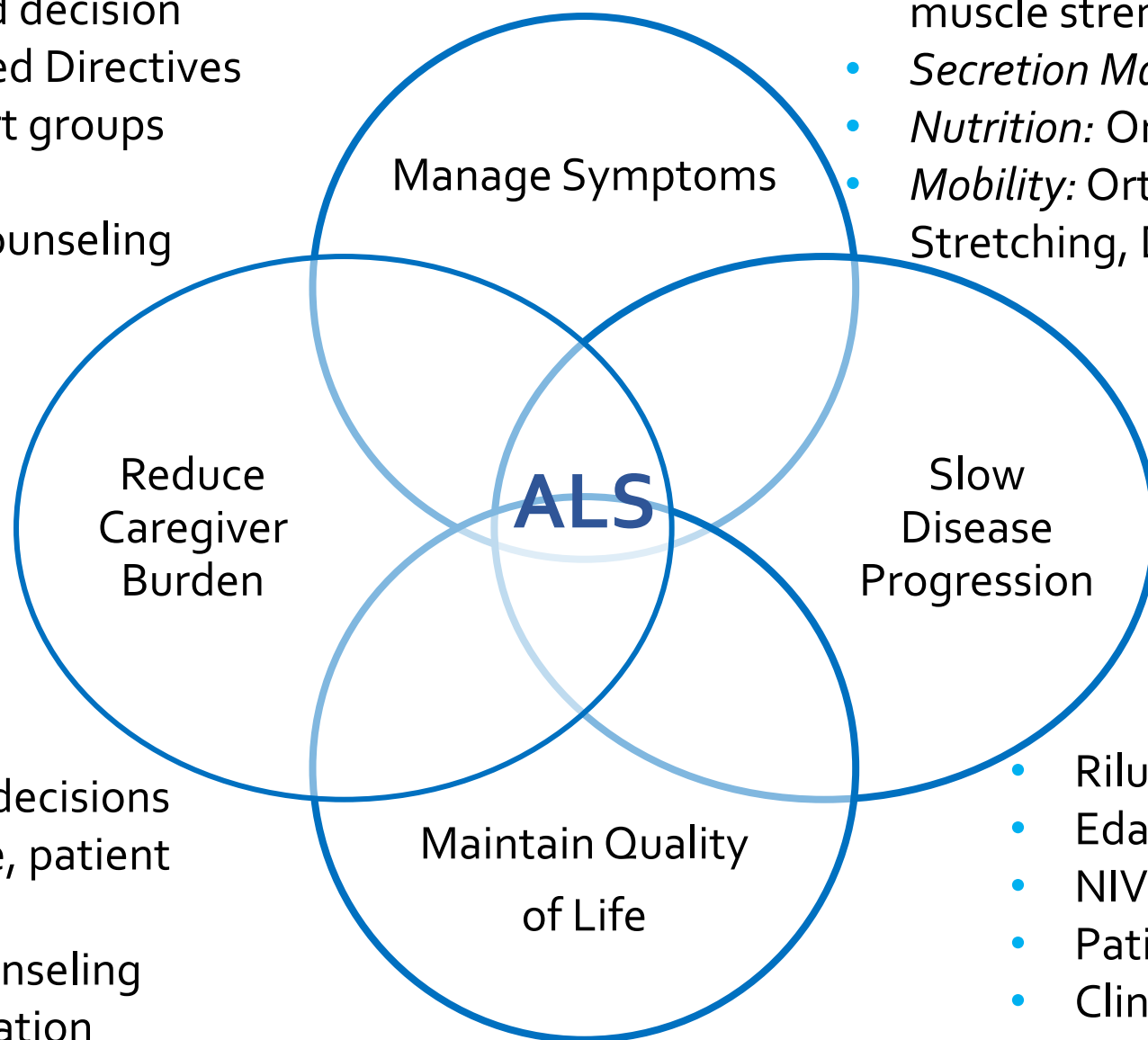
A word cloud centered around the word "overwhelming". The word "overwhelming" is the largest and most prominent, rendered in a dark blue font. Surrounding it are numerous other words in various sizes and colors (red, orange, yellow, green, blue, and purple). The words are arranged in a circular pattern, with some overlapping. The words include: uncontrollable, very'large, preponderance, triumvirate, irrepressible, overpowering, prodigious, hulking, inordinate, unbearable, irrefutable, devastating, huge, most, unusually, sweeping, fantastic, compelling, forceful, personage, massive, big, consuming, bulk, bulky, immense, untold, preeminence, lot, much, expanded, large, enormous, vast, tremendous, gigantic, dominance, gladness, profound, profuse, intense, formidable, irresistible, very'strong, upstream, majority, incalculable, oppressive, stupendous, voluminous, staggering, shattering, unwieldy, and unutterable.

overwhelming

Goals of Multidisciplinary ALS Clinic Visits



- Life planning and decision making, Advanced Directives
- Access to support groups
- Respite care
- Mental Health counseling



- *Rx*: Nuedexta, baclofen, pain & sleep meds
- *Respiratory (early)*: Breath stacking/Respiratory muscle strength training
- *Secretion Management*: rx, botox, radiation
- *Nutrition*: Oral supplements , PEG
- *Mobility*: Orthotics, Botox, Passive ROM/ Stretching, DME

- Life planning and decisions
- Individualized care, patient navigates journey
- Mental Health counseling
- Resource Identification
- Support groups

- Riluzole (+ ~89 days to NIVV/IV)
- Edaravone (33% slowing in subgroup)
- NIVV/cough assist
- Patient and caregiver education
- Clinical Trials

ALS: Nuts and Bolts

- Important Disease Characteristics influencing care:
 - Onset Type (where symptoms began)
 - Body Mass Index (BMI)
 - Degree of respiratory involvement
 - Family History:
 - ALS, Motor neuron disease
 - Psychiatric illness, dementia

Impact on Life and Function

Breathing



Swallowing



Quality of Life



Cognition



Mobility



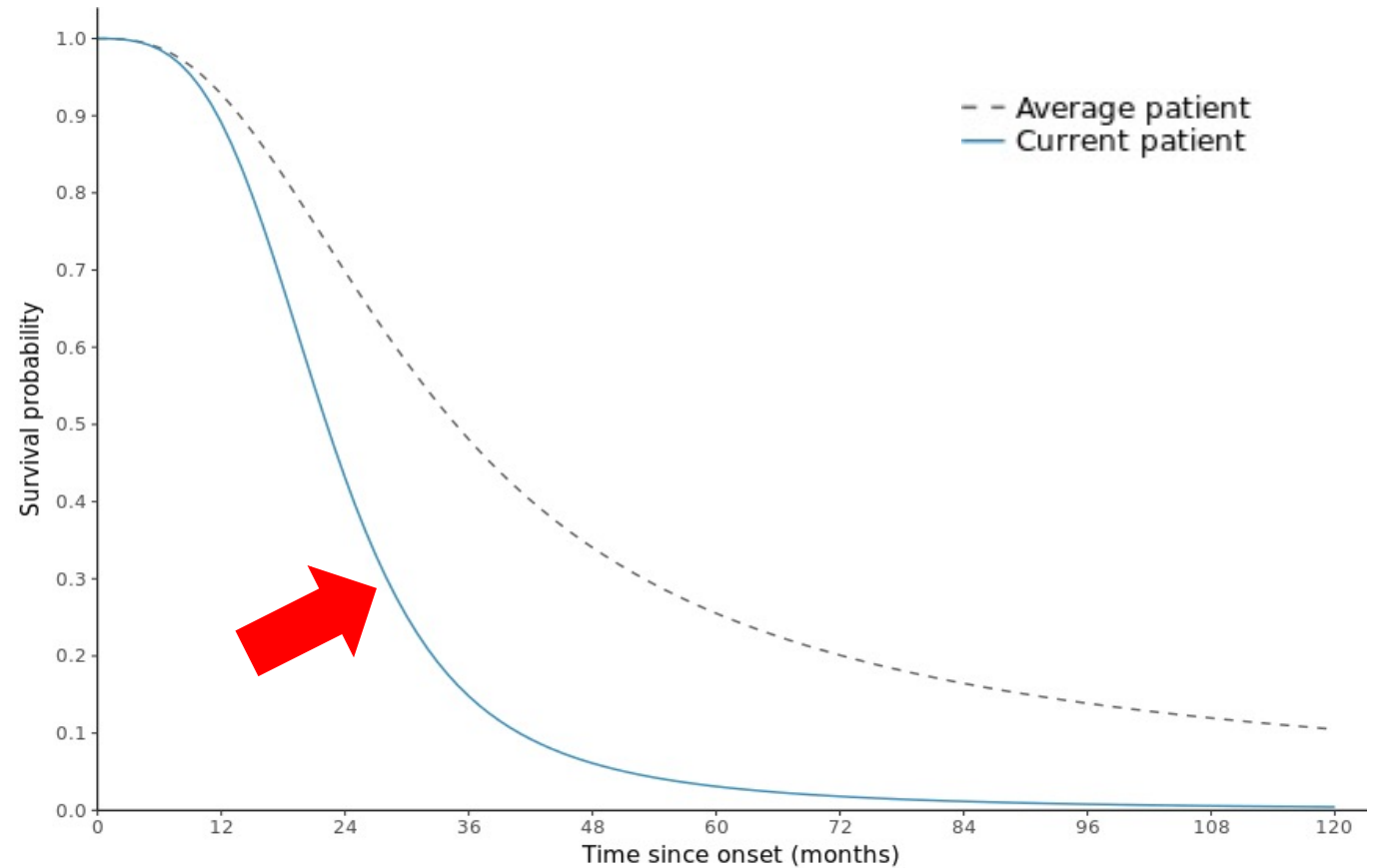
Communication



ALS progresses at different rates

Patient Demographics

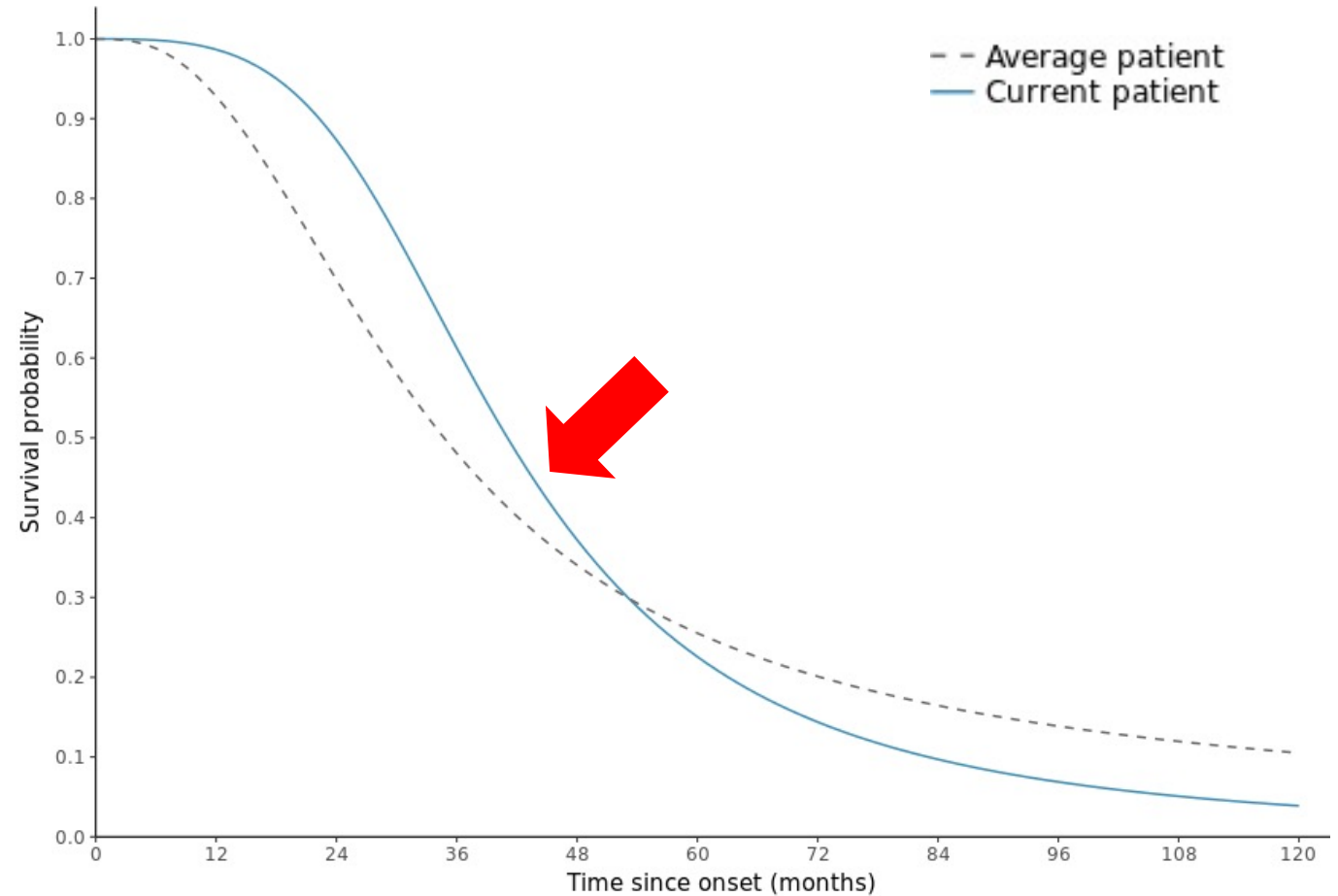
Predictors	Data
Age at onset (years)	66.0
Diagnostic delay (months)	9.9
Progression rate (points decrease on ALSFRS-R per month)	1.22
Forced vital capacity (% of predicted)	65
Definite ALS (according to El Escorial criteria)	Yes
Frontotemporal dementia	Absent
C9orf72 repeat expansion	Absent
Site of onset	Bulbar



ALS progresses at different rates

Patient Demographics

Predictors	Data
Age at onset (years)	64.7
Diagnostic delay (months)	13.8
Progression rate (points decrease on ALSFRS-R per month)	0.29
Forced vital capacity (% of predicted)	75
Definite ALS (according to El Escorial criteria)	Yes
Frontotemporal dementia	Absent
C9orf72 repeat expansion	Absent
Site of onset	Spinal



A Day in ALS Clinic

Neuromuscular Disease Specialist (MD)

- Assesses:
 - Muscle twitching
 - Muscle cramping
 - Muscle strength
 - Reflexes

- Diagnosis
- Head to toe evaluation
- Puts the puzzle pieces together
- Medication management
- Advanced Care Planning



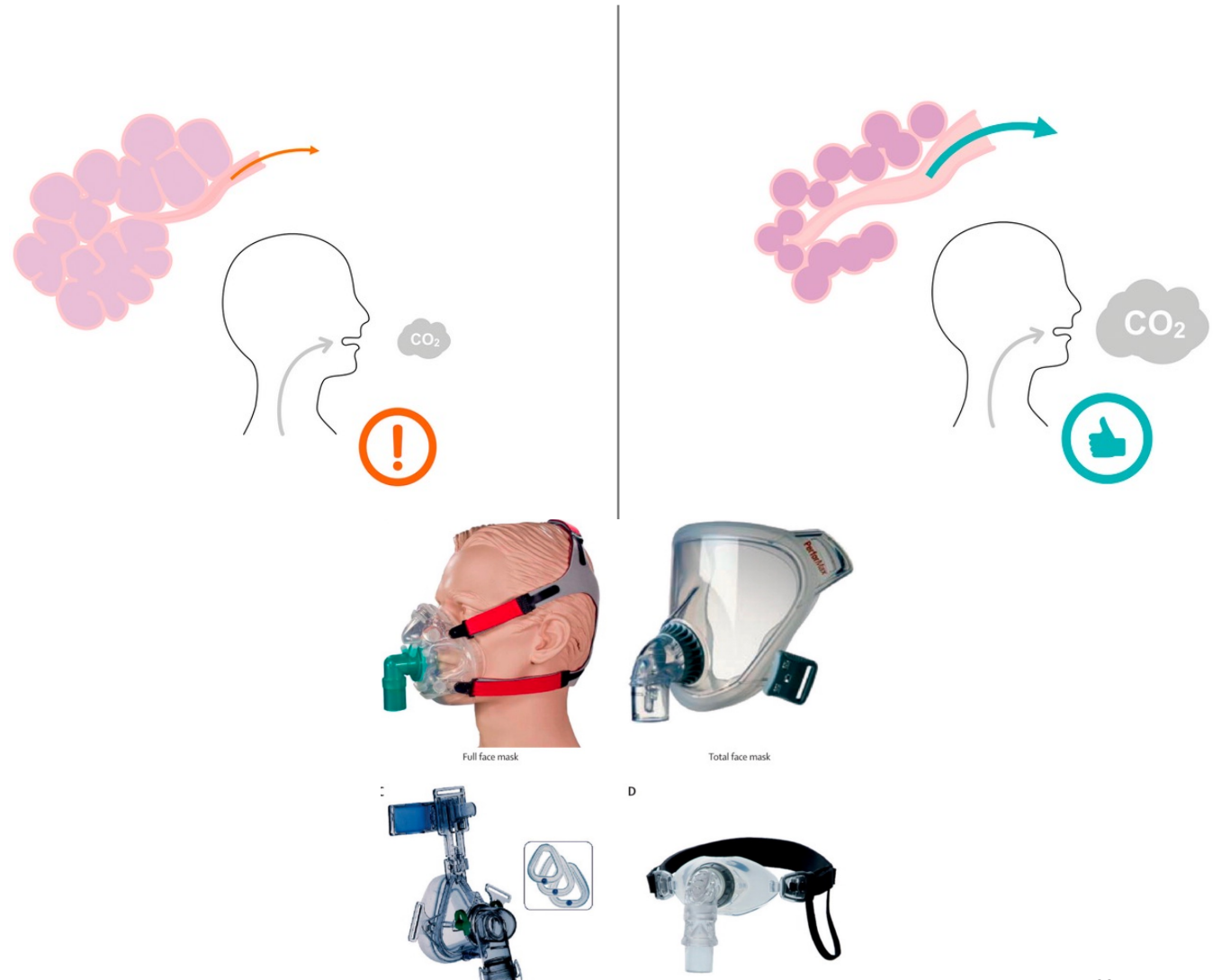
Treatment ▾	Podcast	Mechanism ▾	Pre-Clinical ▾	Cases ▾	Trials ▾	Risks ▾
Ketogenic Diets (2021)		B	C	D	U	D
Vitamin C (2021)		C	C	B	F	B
Melatonin (2021)		A	C	B	U	B

Supplements and Alternative Off-Label Treatments

Respiratory Management

Shortness of Breath

NIVV: Noninvasive Volume Ventilation



Respiratory Management

- Improve weak Cough
- Aide in managing secretions
- Prevent respiratory infections

Weak Cough/ Managing Secretions Cough Augmentation

Cough Assist Device



- Treatment protocol:
- 3-5 coughs
- Performed 3x /day

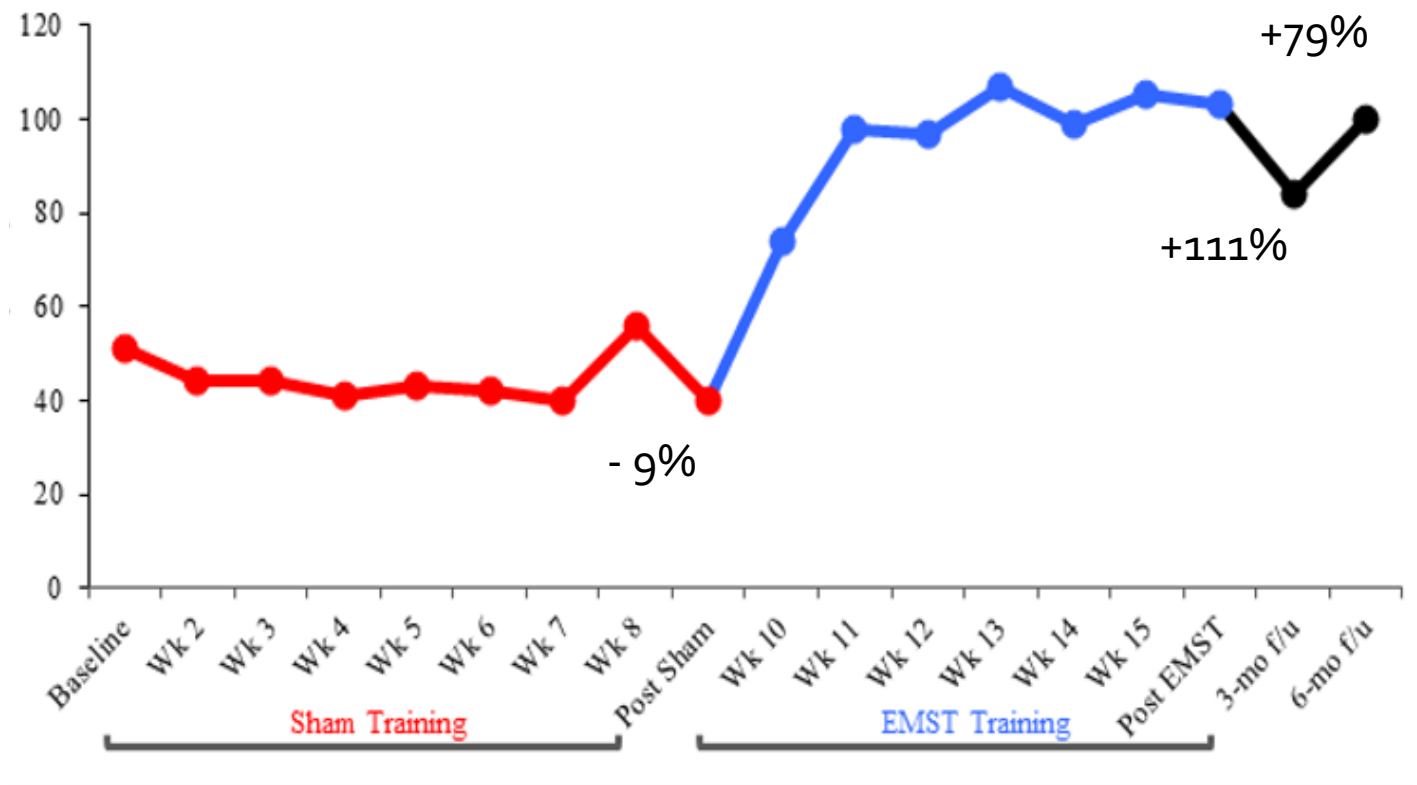
Oral Suction Device

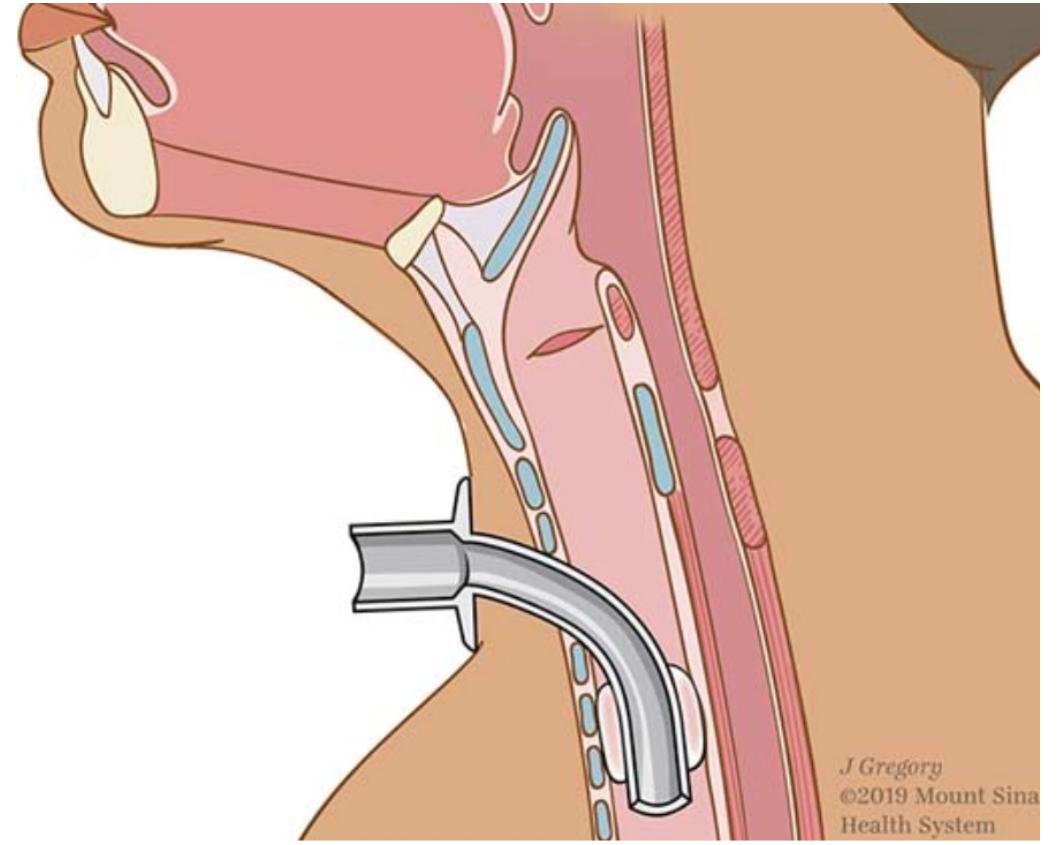
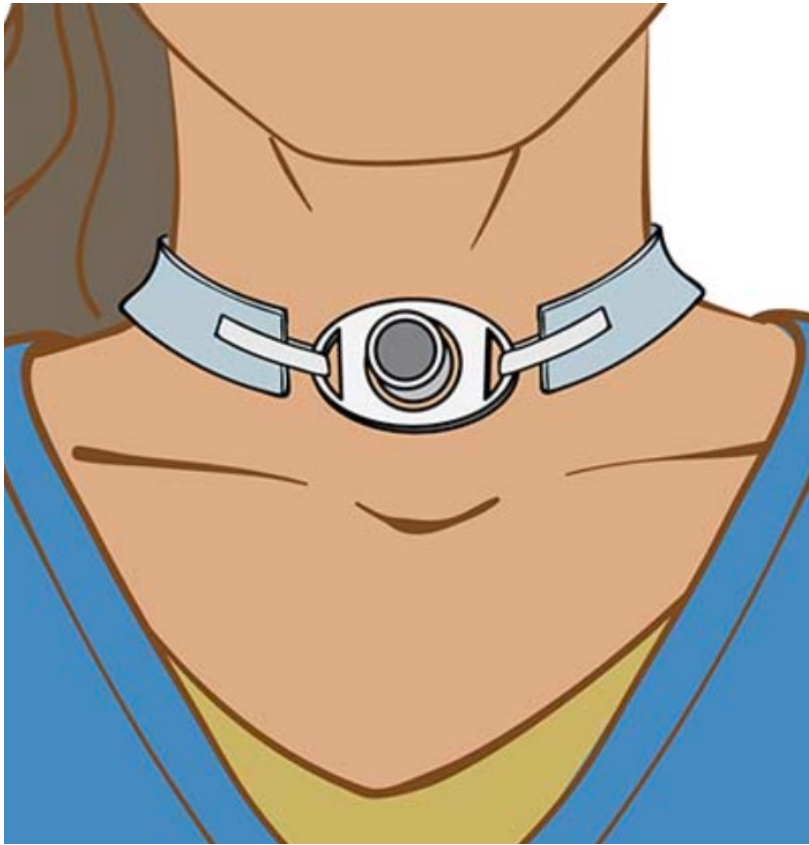


- Use as needed
- Can be combined with medication:
 - Atropine drops
 - Scopolamine patch
 - Botox
 - Various other meds.

Respiratory: Cough Augmentation

Respiratory Muscle Strength Training





Invasive Ventilation: Tracheostomy

Respiratory Support when NIVV is no longer supportive and/or secretions unmanageable

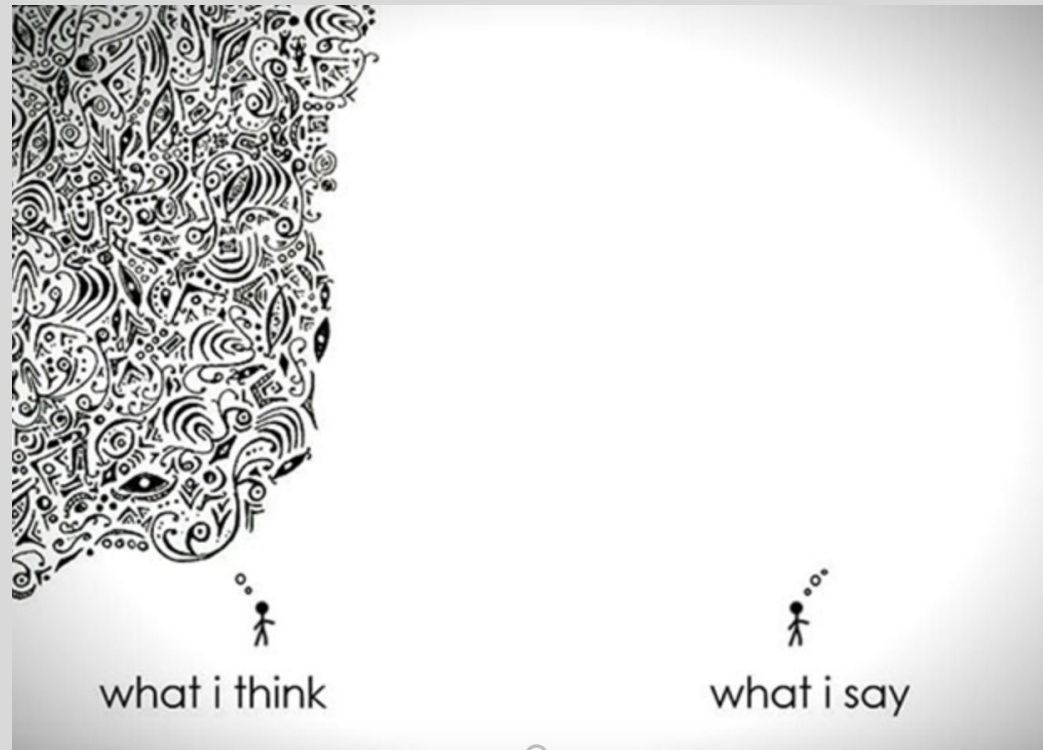
Speech Language Pathologist (SLP)

Swallowing difficulty (dysphagia) & Airway Protection



Speech-Language
Pathologist (SLP)

Communication
Evaluation



Voice Preservation & Communication Strategies

Voice & Message Banking

Compensatory Strategies, Energy Conservation

Alternative and Augmentative Communication (AAC)

Low-tech Communication

High-tech Communication

- Body Mass Index (BMI) a significant predictor of disease progression and survival
- Low BMI associated with faster progression

Dietitian

- Nutrition Support and Management
- Feeding Tube Placement

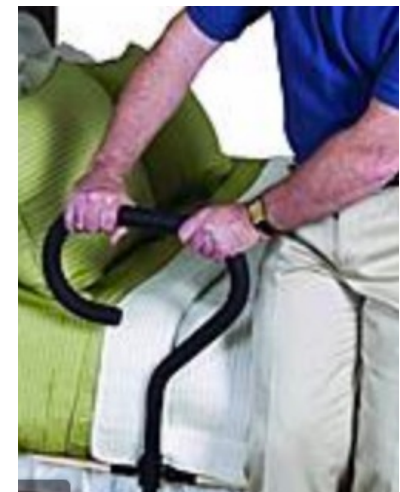
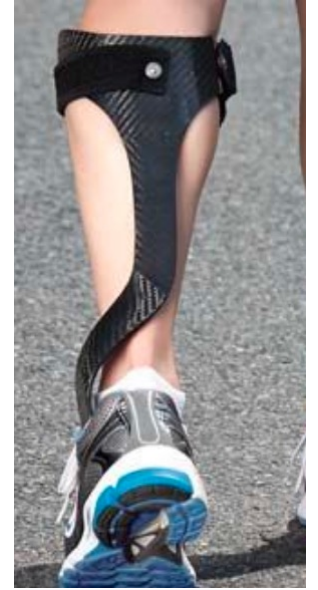
Common Recommendations

- Smaller, more frequent meals
- Snack often
- High calorie sauces, condiment, EVOO
- Benecalorie, oral supplements
- Feeding tube placement

Physical and Occupational Therapy

“DME”: Durable Medical Equipment

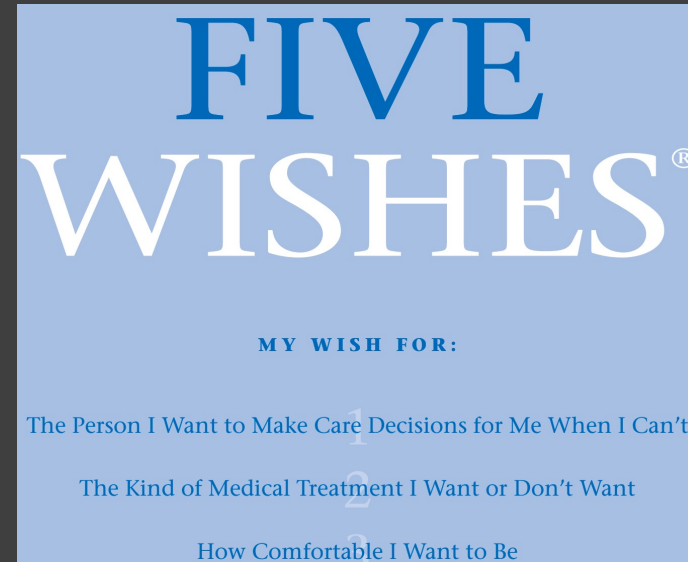
Home Exercise Programs



Social Worker,
ALSA Liaison

(aka: The Glue)

- The Resource Gurus
- Insurance, home health, DME

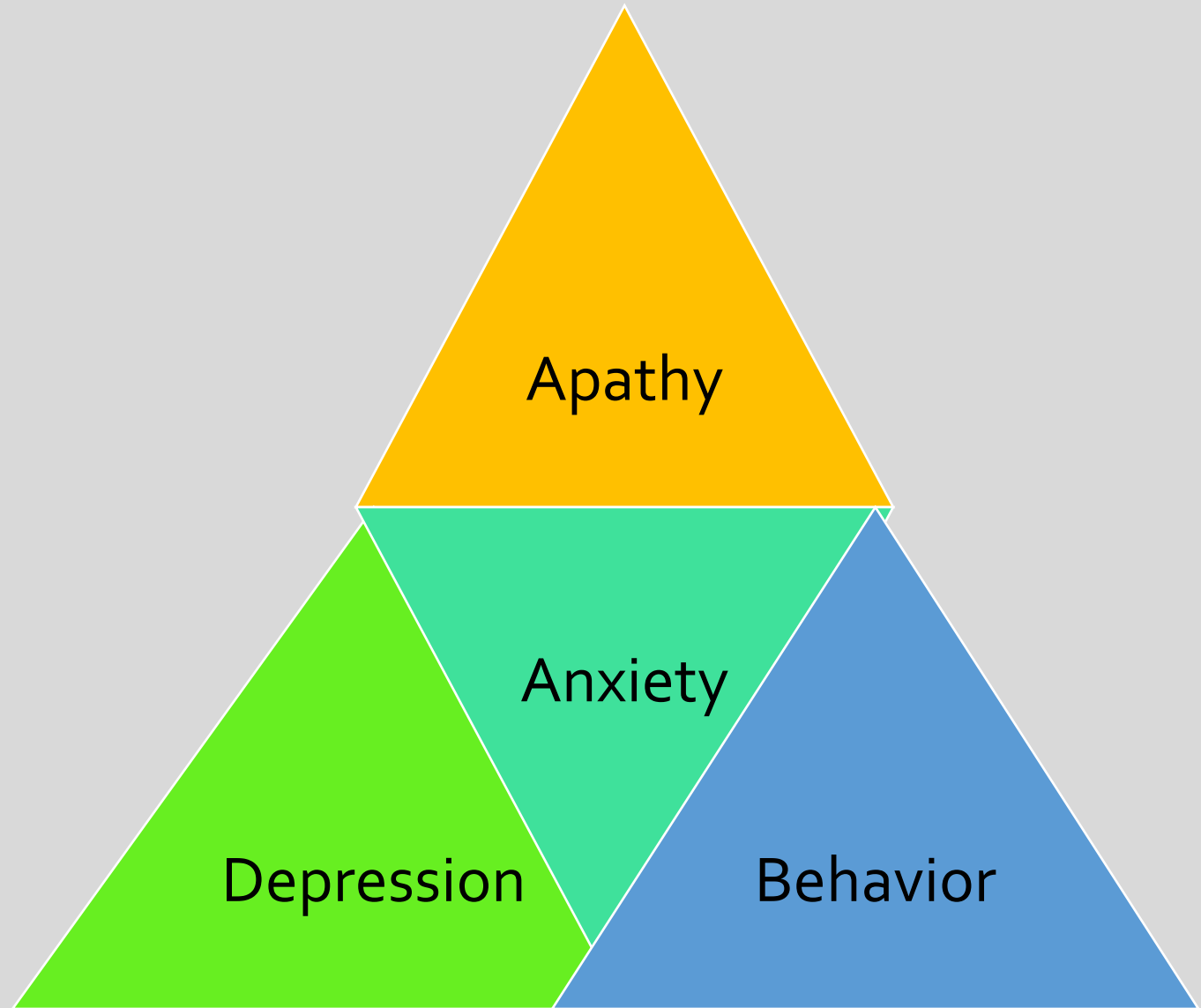


ALSA Registry

<https://www.als.org/advocacy/als-registry>

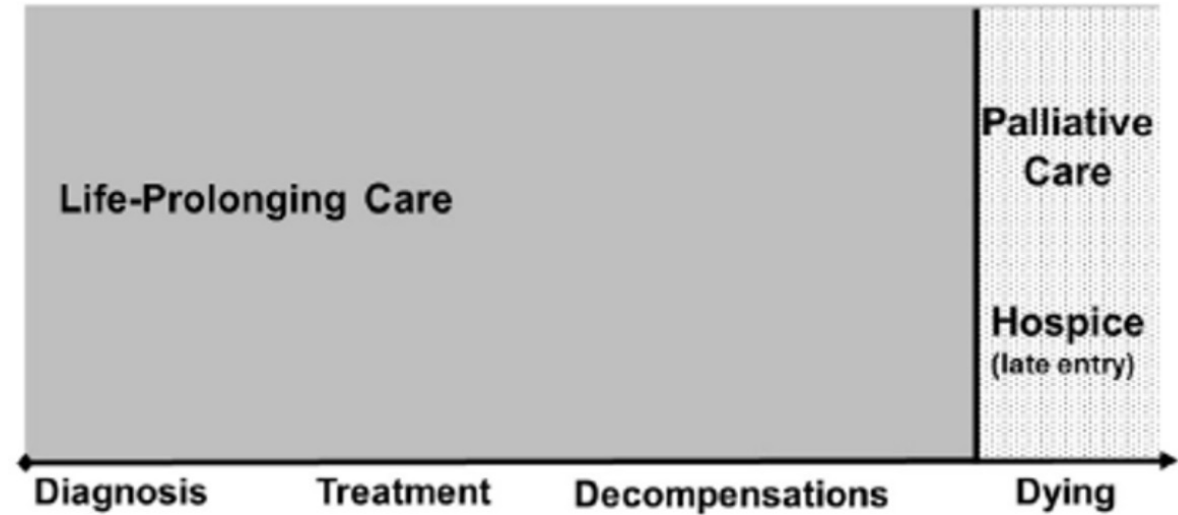
Neuropsychology

Address the emotional, cognitive and quality of life issues that arise when confronting the challenges of living with ALS

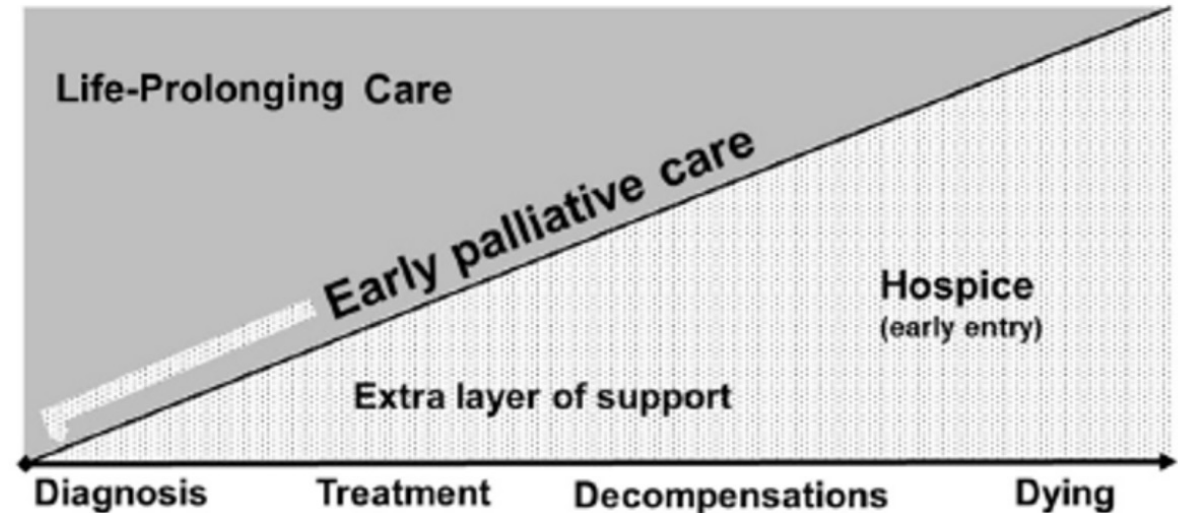


Palliative Care

Extra layer of support and care throughout the duration of disease



Current Paradigm: Disease Trajectory with Late Palliative Care Intervention



Palliative Care

Helpful at *any time* throughout the disease for pALS and cALS



Multidisciplinary Care Team Approach

Specialist	Role
Neurology/ NP	Diagnosis, Medication management
SLP	Swallow, Airway protection, Communication
OT/PT	Fine motor skills (feeding), mobility, home exercise program
Respiratory	Cough Augmentation, Respiratory therapy
Social Work	Service provision and setup, Insurance, Resources
Dietitian	Nutrition Management
Nurse	Vitals, Clinic coordination, Patient Advocacy
Palliative Care	Additional support layer, Life decision-making, Hospice consult
Neuropsychology	Emotion and well-being, changes in thinking, behavior

Clinical Research Opportunities

Interventional



Observational



Biobank



Goals of ALS Care

Comprehensive,
coordinated care: family,
caregivers, friends, ALS
clinic team

Proactive intervention vs.
Reactive intervention

- Energy Conservation
- Prophylactic decision-making
- Patient Education

Summary

Multidisciplinary clinics can improve QOL, symptom management, disease-related complications and survival

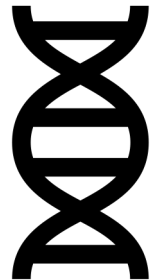


Empowering pALS and cALS through research design and participation

Clinical Research Learning Institute® (CRLI)



Visits typically occur every 3-6 months and last for 3.5 hours to see multiple providers



“We’ve been wrong about what our job is in medicine. We think our job is to ensure health and survival. But really it is larger than that. It is to enable well-being.”

-Dr. Atul Gawande, Being Mortal: Medicine and What Matters in the End

Thank you

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