



Improving Quality of Life and Care in Lennox-Gastaut Syndrome: Best Practices for Collaborative, Family- and Patient-Centered Management

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**LEARNING
OBJECTIVE 1**

Identify non-seizure features and comorbidities that may impact patients with LGS.



**LEARNING
OBJECTIVE 2**

Integrate strategies to address QoL concerns of patients with LGS and their caregivers.



**LEARNING
OBJECTIVE** **3**

Incorporate best practices for long-term planning as part of the transition from pediatric to adult care for patients with LGS and their caregivers, including team-based, interprofessional care.

Challenges in Identifying Non-Seizure Features of LGS



Audience Response



? Which of the following non-seizure symptoms seen in LGS do you consider the most burdensome?

- A. Cognitive impairment
- B. Behavior issues
- C. Feeding difficulties
- D. Mobility issues

Lennox-Gastaut Syndrome

Developmental epileptic encephalopathy (DEE) characterized by treatment-resistant seizures and cognitive and behavioral abnormalities

Accounts for 1%-2% of all childhood epilepsies

Often evolves from other infantile epilepsy syndromes such as infantile epileptic spasms syndrome (IESS)/West syndrome

Typically presents between 18 months and 8 years of age with peak onset between 3 to 5 years

Persists into adulthood in nearly all cases

Different Causes of LGS



Brain injury

Trauma

Birth complications

Infection



Unknown!

Genetic

Metabolic

Brain malformations

Neurocutaneous disorders: NF1 or tuberous sclerosis

NF1 = neurofibromatosis 1.

Asadi-Pooya AA. *Neurol Sci.* 2018;39(3):403-414. Goldsmith IL, et al. *Epilepsia.* 2000;41(4):395-399.

LGS Diagnostic Triad



Multiple Seizure Types

- Tonic,* atypical absence, tonic and atonic drop attacks, myoclonic, focal, non-convulsive status epilepticus

Abnormal EEG

- Interictal slow spike waves (< 2.5 Hz)
- Paroxysmal fast rhythms (10-20 Hz) that are mainly during non-REM sleep

Cognitive Impairment

- Spectrum of cognitive challenges from learning disabilities to intellectual disability

*Required for diagnosis.

EEG = electroencephalogram; REM = rapid eye movement.

Specchio N, et al. *Epilepsia*. 2022;63(6):1398-1442.

Seizures in LGS



Diagnosis requires tonic seizures and at least one other seizure type

Hallmark Seizures of LGS:

- **Tonic** (predominantly at night): required for diagnosis
- Atonic
- Atypical absence

Other Associated Seizure Types:

- Epileptic spasms
- Non-convulsive status epilepticus
- Focal impaired awareness
- Generalized tonic-clonic
- Myoclonic
- Atonic
- Atypical absence

Non-Seizure Symptoms (NSS): Cognitive

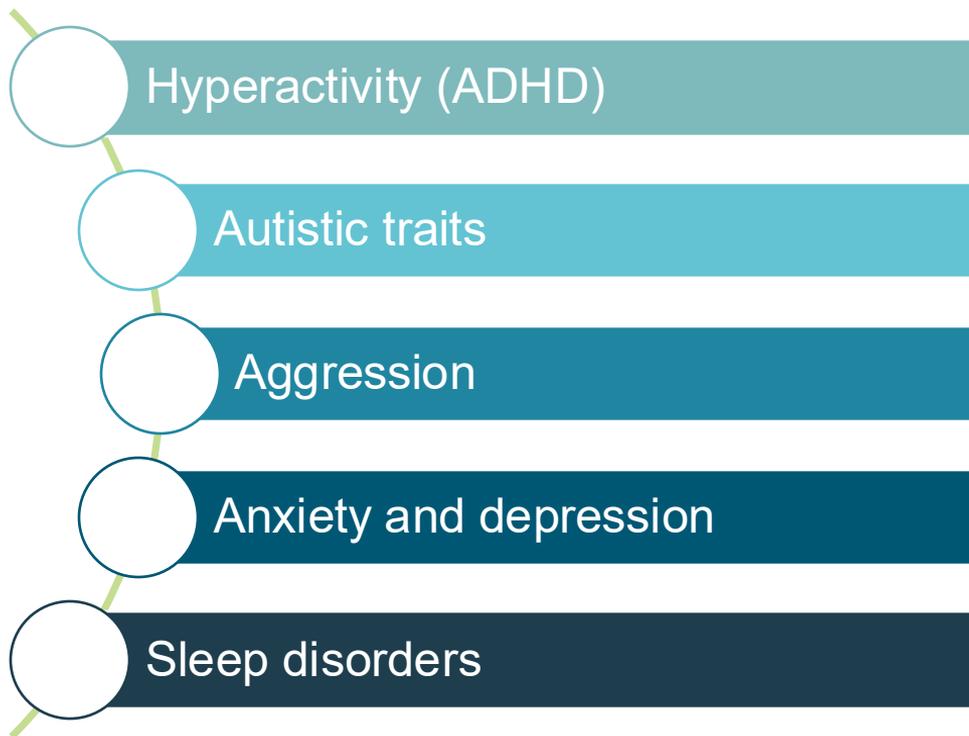
Intellectual disability occurs in 75%-95% of people with LGS

Over time, there is developmental slowing, plateauing, or regression

Impairment is primarily related to the cause of the seizures

Seizure burden and seizure treatments can further impair cognition and learning

NSS: Mood and Behavioral



Behavior issues often stem from frustration due to an inability to communicate or understand what is happening; the intensity of behavioral outbursts has greater consequences as the patient ages

ADHD = attention-deficit/hyperactivity disorder.

Marchese F, et al. *Sn Compr Clin Med*. 2021;3:2167-2179. Glauser TA. *Epilepsia*. 2004;45:23-26. Camfield PR, et al. *Epilepsia*. 2011;52:21-17.

NSS: Motor and Mobility

- Staggered gait and difficulty walking that **increases** with age
- Many individuals **require mobility support** such as wheelchairs or walkers
- Motor issues contribute to dysphagia and **challenges** in dressing and caring for self
- Motor problems may **worsen** with age



NSS: Mortality



Nearly 14-fold increase in death compared to peers

SUDEP is the main contributor to early mortality

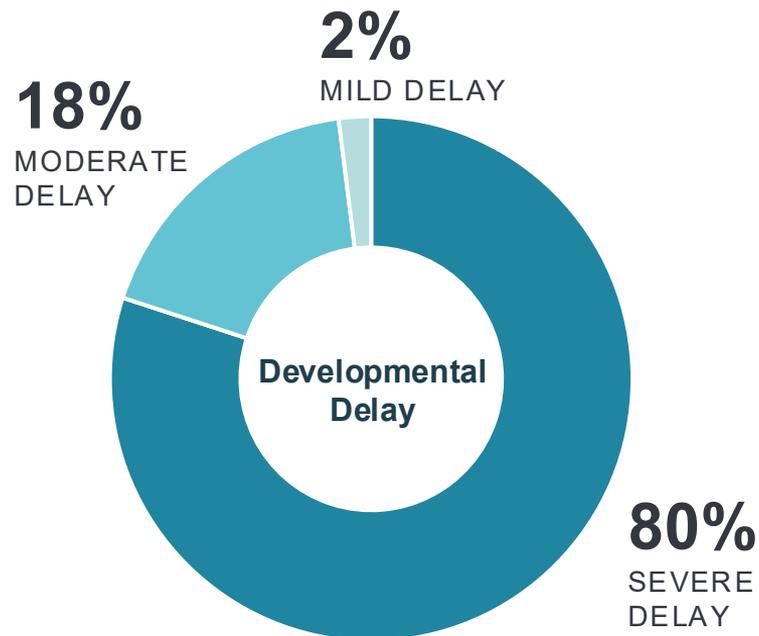
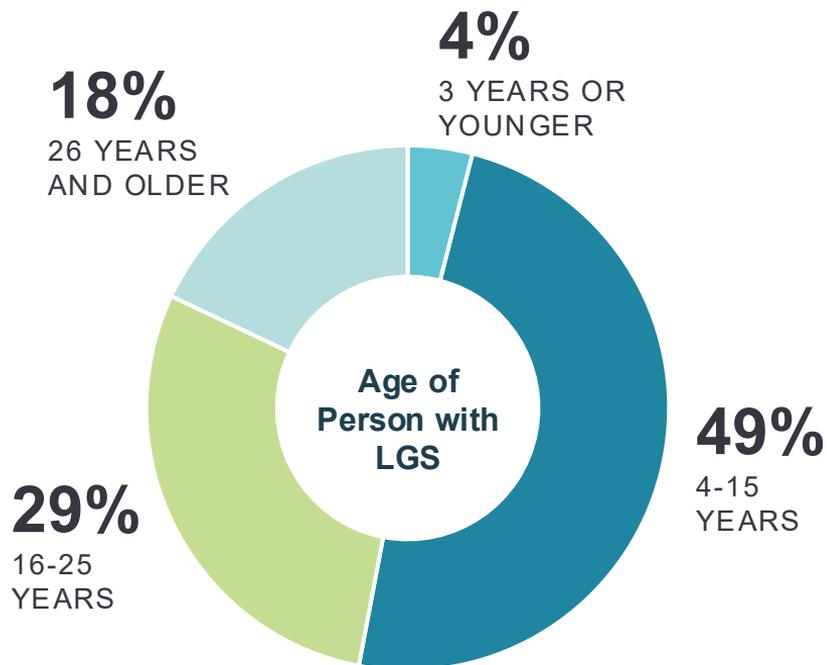
True extent of mortality may be underestimated due to underdiagnosis

More Than Just Seizures

*Addressing Quality of Life for Patients
with LGS Transitioning to Adulthood*

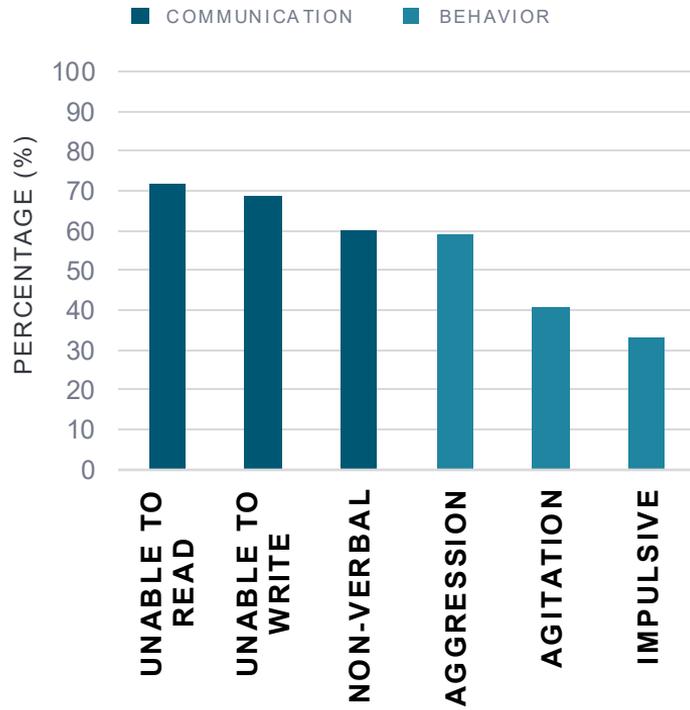


2018 LGS Foundation Caregiver Survey

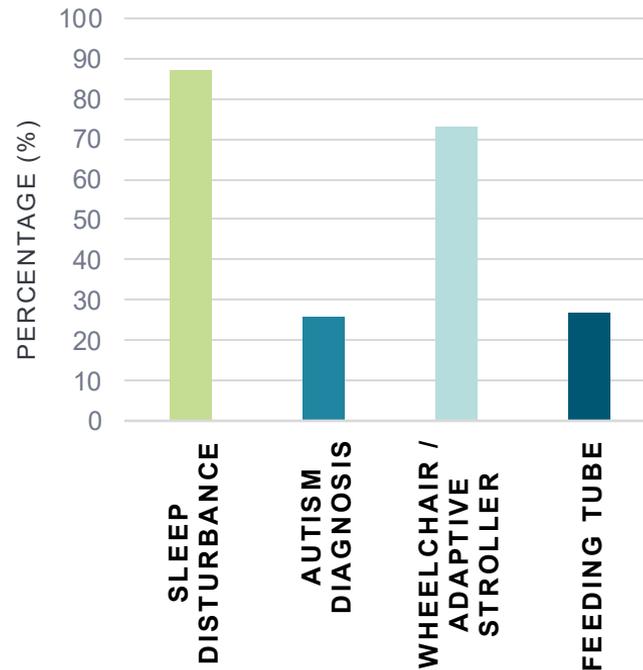


2018 LGS Foundation Caregiver Survey

Communication and Behavior



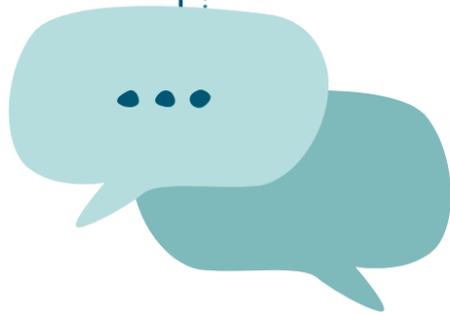
Sleep, Autism, Mobility, and Feeding



Medical Complexity Beyond Seizures



Video Clip 1



Impact on Patient QoL

Cognitive/Behavioral

- Limited ability to communicate
- Frustration, aggression



Physical

- Risk of falls with seizures
- Need for a wheelchair



Social

- Limited social interactions
- Limited recreational activities



Mortality

- Increased mortality compared to other children



Impact on Caregiver/Family QoL



Implications and Consequences

Inability to make plans

Managing complex medical care (appointments, specialists, medications, etc.)

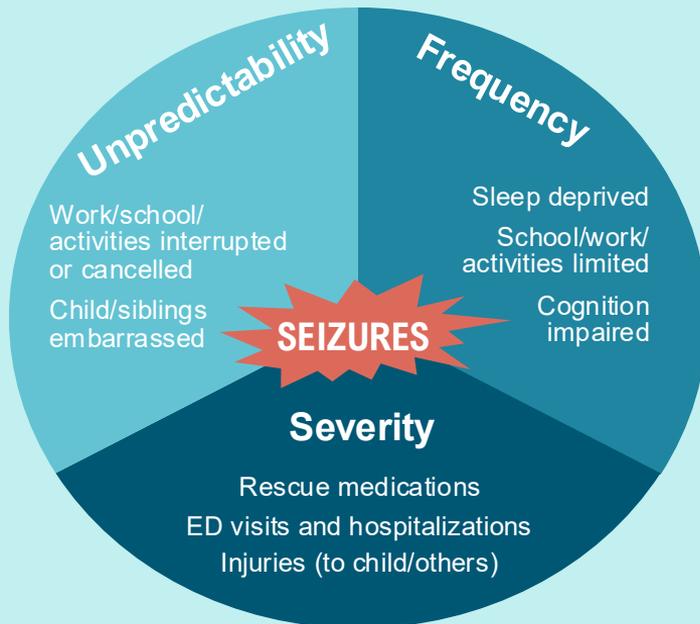
Battling system-level barriers (insurance, finances, advocacy)

Educating self, family, outside caregivers (school, home nurse)

Balancing impacts/needs of other family members

Stranger in home (home nurse)

Monitoring seizures, behavior, cognition, sleep (SUDEP), diet



Constant stress overload

Constant fear/anxiety/worry

Self-sacrifice and guilt about not doing enough

Chronic sleep deprivation, 24/7 exhaustion

Wear on relationships (marriage, siblings, friends)

Isolation and depression

Loss of hope

Fear for the future

“Surviving, not thriving”

Chronic Traumatic Stress

ED = emergency department.

Berg AT, et al. *Epilepsia Open*. 2019;4:293-301. Bailey LD, et al. *Epilepsy Behav*. 2020;112:107377.

Two Caregiver Perspectives: Early-Stage vs Long-Term Needs



Caregiver priorities shift over time – from seizure control to long-term quality of life

Caregivers of **younger** patients:

- Often in “fight or flight” mode – overwhelmed and isolated
- Emergency-focused: fear of death; frequently in ED
- Unsure how to access or trust support systems

Caregivers of **older** patients:

- Focus shifts to adult care settings, inclusion, and long-term QoL
- Worry about aging caregivers’ declining health and ability to continue providing care
- Burnout, fatigue, and helplessness more common

“ We want fun activities and a college life for our child – not just babysitting.

– **Caregiver** of a young adult with LGS

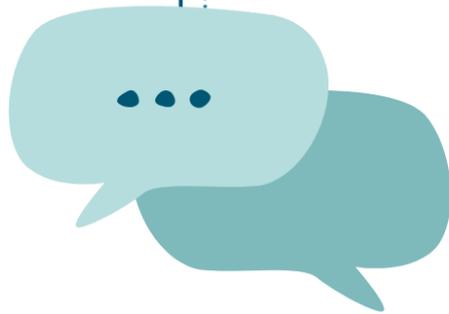
”
Reflect on how younger vs older caregiver needs differ, and how clinical messaging can be adapted.

Education should meet caregivers where they are in this emotional journey

The Ongoing Learning Curve for Caregivers

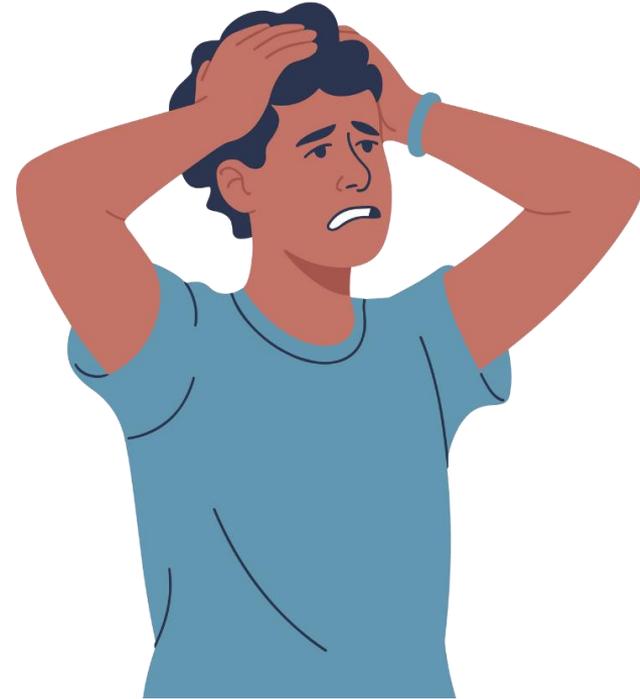


Video Clip 2



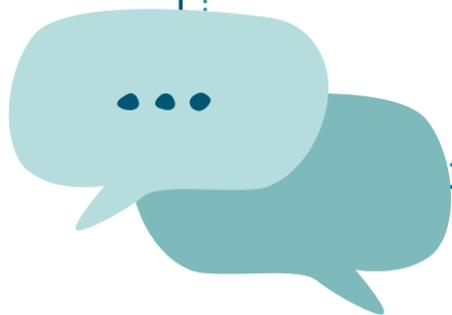
Caregiver Feedback: What HCPs Should Know – Early Barriers and Engagement

- Many families say clinicians **sugarcoat the diagnosis**
- Additional **follow-up or take-home resources** are necessary to absorb LGS education
- Emergency care often feels **unsupportive or dismissive**; ED visits rarely result in follow-up communication with the child's neurologist or care team
- **Seizure plans** should clarify team roles and next steps
- Families **do not know where to start** and fear **rejection or failure** when seeking peer support and other services
- Early success in finding resources **encourages engagement** and builds confidence



Caregiver Voice: Navigating Emergency Care

Video Clip 3



Caregiver Feedback: What HCPs Should Know – Lifelong Systems and Support

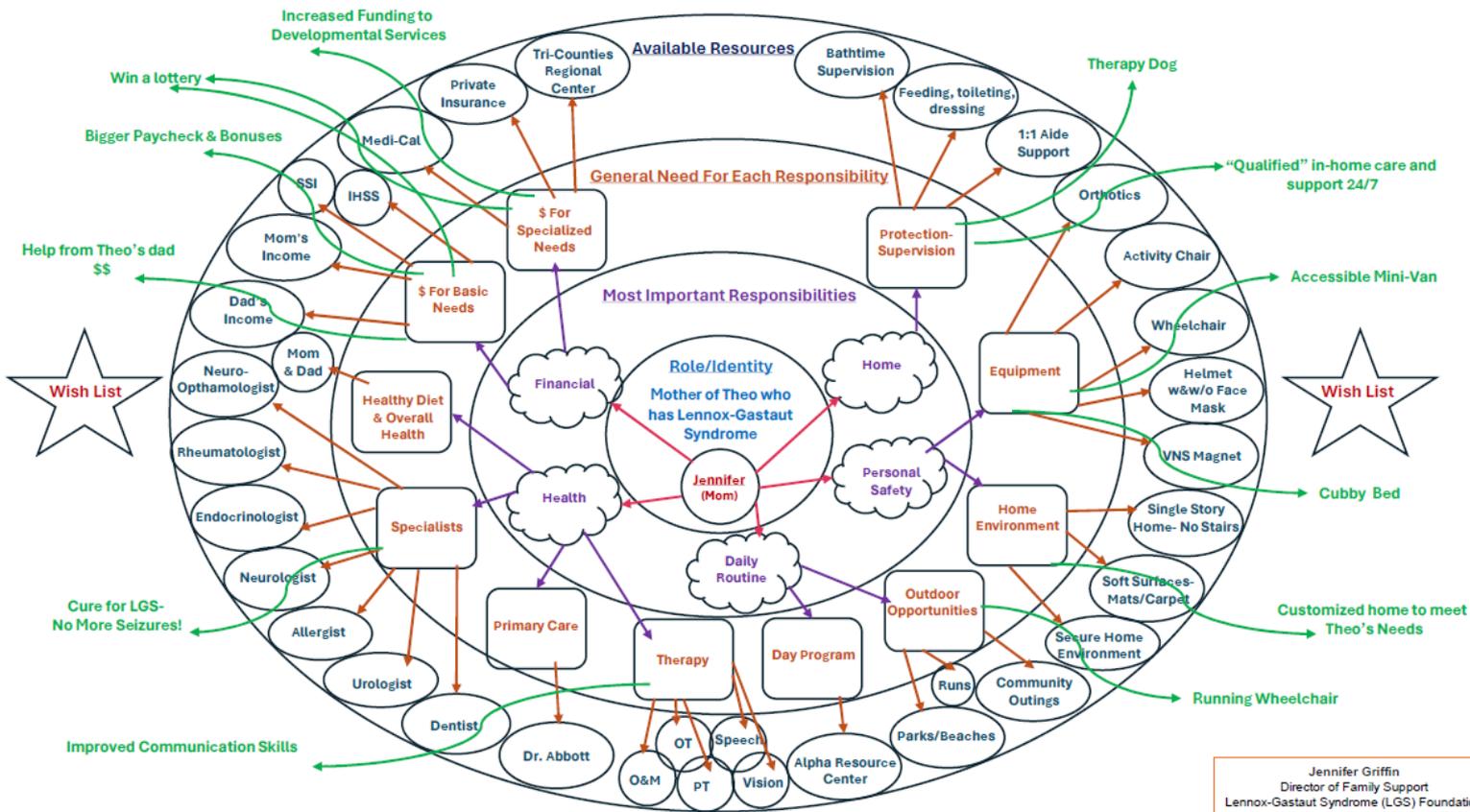
- Families feel underrepresented in care and **judged by schools for “not disciplining”** LGS-related behaviors
- Seizure **stigma excludes families** from programs despite meeting functional criteria
- Aging caregivers report **isolation and fear** that ongoing system failures will discourage continued engagement
- Families report that **group homes often fail** to meet the complex needs of individuals with LGS

I don't know how ready I still am to even talk about long-term planning.

– **Caregiver** of a young adult with LGS

HCPs should consider how referral and planning conversations reflect real caregiver experiences.

Burden of LGS on the Caregiver



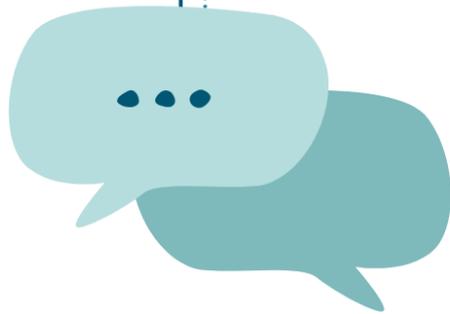
IHSS = in-home supportive services; O&M = orientation and mobility; OT = occupational therapy; PT = physical therapy; SSI = supplemental security income.

Slide courtesy of Jennifer Griffin.

Accepting Support as a Caregiver



Video Clip 4





Caring for the Caregiver

Faculty Panel Discussion



Audience Response

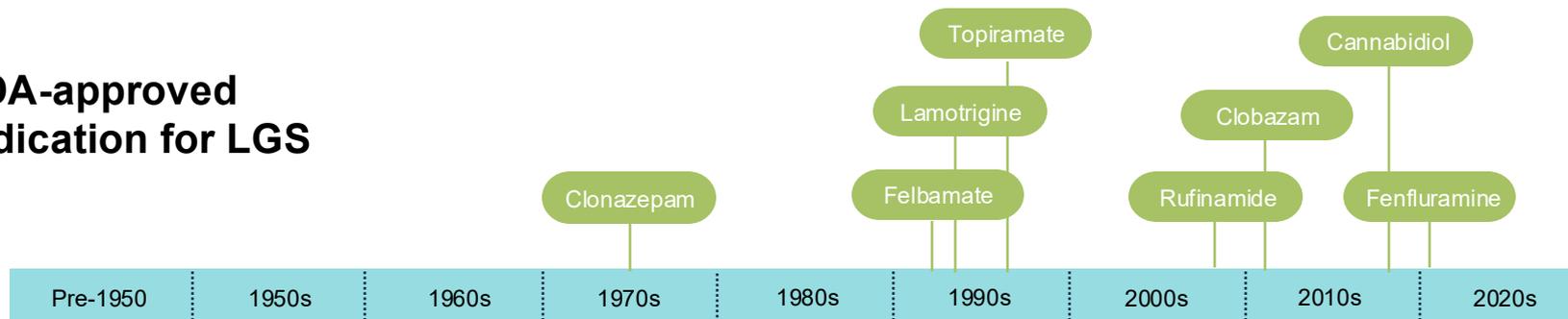


? What is your main goal when you treat individuals with LGS?

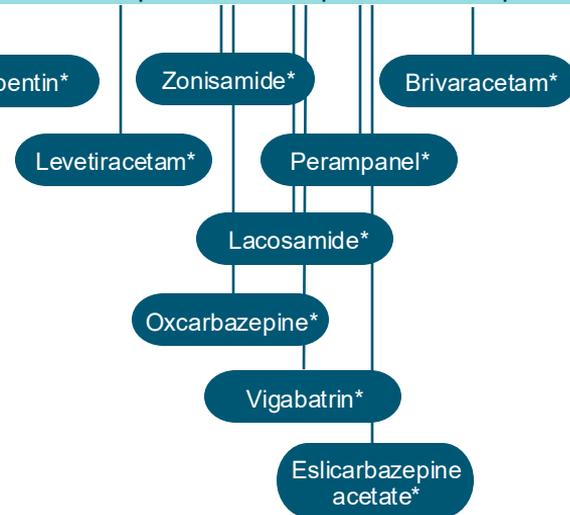
- A. Achieve seizure freedom
- B. Improve patient motor skills
- C. Address behavior and sleep issues
- D. Reduce seizure frequency and severity
- E. Minimize treatment side effects

Evolution of Medication for LGS

FDA-approved indication for LGS



No FDA indication for LGS



*These agents are not indicated for the treatment of LGS.
Sankar R, et al. *Seizure*. 2023;110:42-57.

Impact of ASMs on QoL and NSS

ASM	Behavior	Mood	Cognition	Sleep
Valproic acid*	Positive	Neutral	Positive	Positive
Clobazam	Positive	Positive	Positive	Neutral
Topiramate*	Negative	Negative	Negative	Positive
Levetiracetam*	Negative	Negative	Neutral	Positive
Perampanel*	Negative	Negative	Positive	Neutral
Ethosuximide*	Positive	Positive	Positive	Negative
Lacosamide*	Positive	Positive	Neutral	Positive
Lamotrigine	Neutral	Neutral	Neutral	Negative
Rufinamide	Positive	Neutral	Neutral	?
Cannabidiol	Positive	Neutral	Neutral	Neutral
Fenfluramine	Positive	Positive	Neutral	?

Positive
Neutral
Negative

*These agents are not indicated for the treatment of LGS.
ASMs = anti-seizure medications.
Strzelczak A, et al. *CNS Drugs*. 2022;36(10):1079-1111.

Safety and Tolerability of ASMs for LGS

Study	Treatment	Any AE, Active/Placebo	Dropouts for Any Reason, Active/Placebo	Dropouts Due to AE, Active/Placebo	Serious AEs, Active/Placebo	Most Common AEs
Ohtsuka et al.	Rufinamide	27/21	4/1	4/1	1/1	Somnolence, pyrexia, decreased appetite, vomiting, status epilepticus
Glaser et al.	Rufinamide	60/52	10/5	9/1	2/2	Somnolence, vomiting, pyrexia, diarrhea
Thiele et al.	Pharmaceutical CBD	74/79	14/1	8/1	20/4	Diarrhea, somnolence, pyrexia, decreased appetite, vomiting
Devinsky et al.	Pharmaceutical CBD	133/55	11/2	7/1	26/7	Somnolence, decreased appetite, diarrhea, URTI, vomiting, pyrexia, nasopharyngitis, status epilepticus
Sachdeo et al.	Topiramate	NA	1/0	NA	11/5	Somnolence, anorexia, nervousness, behavioral problems, fatigue, dizziness, weight loss
Ng et al.	Clobazam	142/40	42/18	29/12	14/2	Somnolence, pyrexia, lethargy, drooling, constipation
Motte et al.	Lamotrigine	NA	3/7	3/7	3/0	Pharyngitis, fever, infection
Felbamate Study Group	Felbamate	NA	1/1	NA	8/3	URTIs, anorexia, vomiting, somnolence, injury, fever, insomnia, nervousness, headache, fatigue, purpura, abnormal gait, ataxia
Knupp et al.	Fenfluramine	78 (0.2 mg/kg) 90 (0.7 mg/kg)/75	--	--	4 (0.2 mg/kg) 11 (0.7 mg/kg)/5	Decreased appetite, somnolence, fatigue

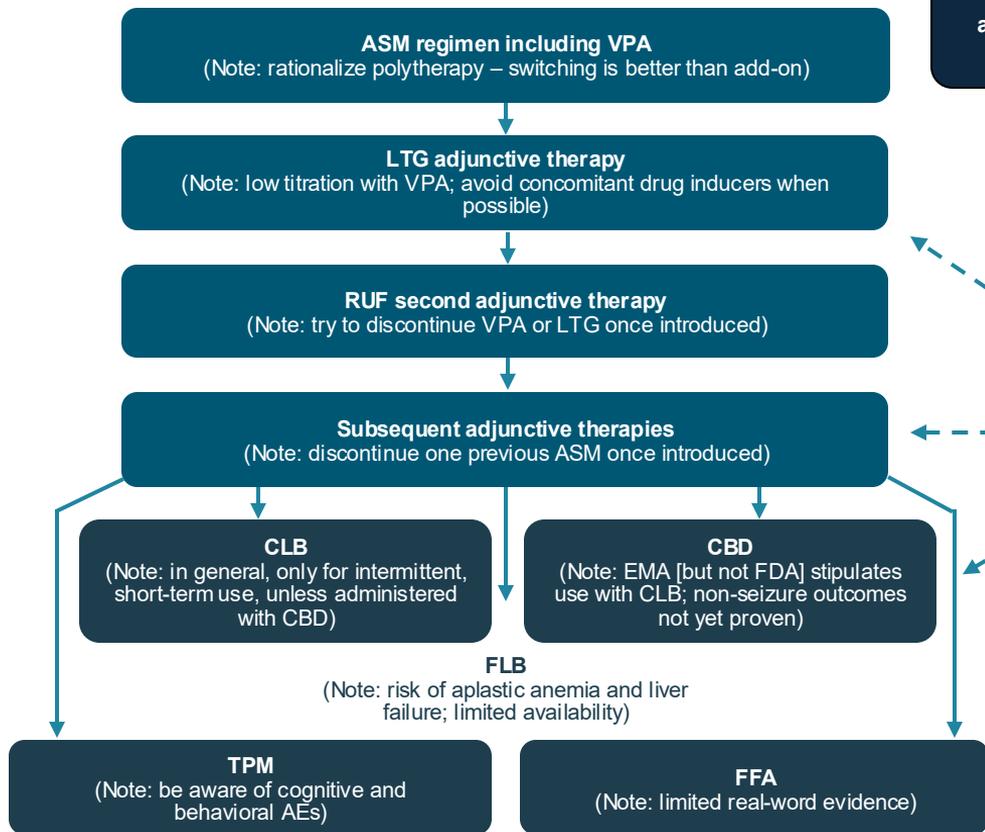
All agents on this slide are FDA-approved for the treatment of LGS.

AE = adverse effect; CBD = cannabidiol; URTI = upper respiratory tract infection.

Zhang L, et al. *Dev Med Child Neurol*. 2022;64(3):305-313. Knupp KG, et al. *JAMA Neurol*. 2022;79(6):554-564.

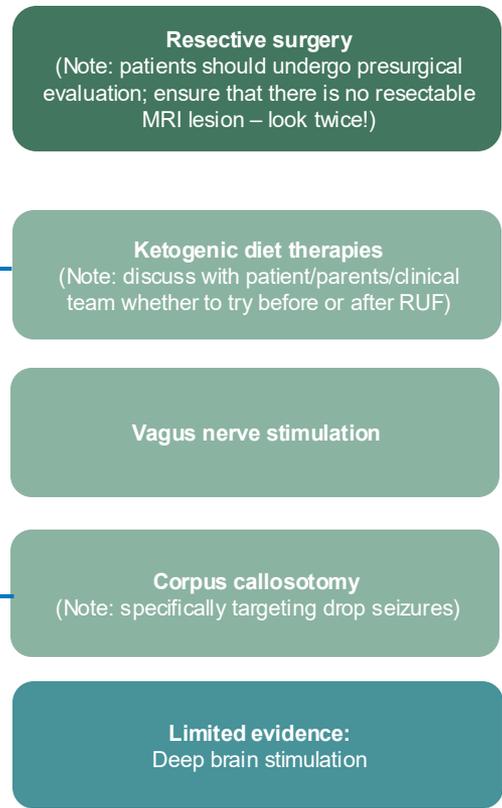
Multimodal Approach to Therapy

Pharmacological Therapy



Personalized approach based on etiology

Non-Pharmacological Therapy



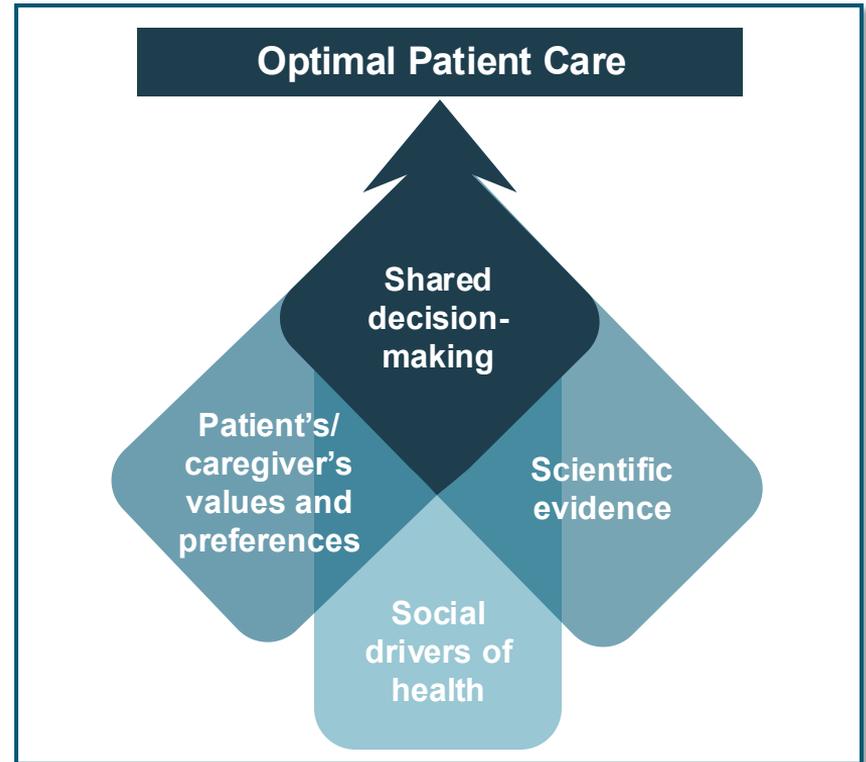
All agents and procedures on this slide are FDA-approved for the treatment of LGS.

CLB = clobazam; EMA = European Medicines Agency; FFA = fenfluramine; FLB = felbamate; LTG = lamotrigine; MRI = magnetic resonance imaging; RUF = rufinamide; TPM = topiramate; VPA = sodium valproate.

Auvin S, et al. *Epilepsia Open*. 2024;10(1):85-106.

Shared Decision-Making in LGS

- Shared decision-making in LGS is done predominantly with the caregiver, but the patient **can and should** be included
- Follow the SHARE model:
 - **S**eek participation
 - **H**elp explore and compare treatment options
 - **A**ssess values and preferences
 - **R**each a decision with patient and caregiver
 - **E**valuate the decision



Supported Decision-Making

- Supported decision-making may be appropriate for patients with LGS, depending on **age** and level of **intellectual impairment**
- Supported decision-making **empowers patients** and helps them to feel they have **agency** in their treatment decisions



**Substitute
Decision-Making**
Someone else makes decisions **on
behalf** of the person



**Supported
Decision-Making**
The person is supported by others to
make more of their **own decisions**



Holistic Approaches to Long-Term Planning for Patients with LGS



Audience Response



? At what age do you generally begin transitioning patients with LGS to adult care?

- A. 12-14 years
- B. 15-17 years
- C. ≥ 18 years
- D. I am not involved in the transition process

Transfer Versus Transition



Health Care Transfer

- Patient's **physical transition** from a child-oriented health care system to an **adult-oriented** health care system
- Involves **new** doctors, locations, insurance, and structure



Health Care Transition

- Patient's **holistic development** as they move from the role of “child patient” to “adult patient”
- **Increased responsibility** for their health, including making appointments, managing prescriptions, and tracking symptoms
- Less reliance on the support of others
- Caregivers often feel unprepared for this shift, especially when support has failed them in the past



Transfer from Pediatric to Adult Care

- Transferring from pediatric to adult care is often **difficult** for both patients and caregivers
- Without planning, families face **delays** in specialty care, poor management of comorbidities, and barriers to adult hospital access
- Adulthood introduces **new challenges**: guardianship, insurance coverage, and long-term care needs
- **Early, structured preparation** improves the likelihood of a smoother and more successful transfer of care
- Unrecognized **medical trauma** may complicate transitions, especially in patients with cognitive or communication barriers

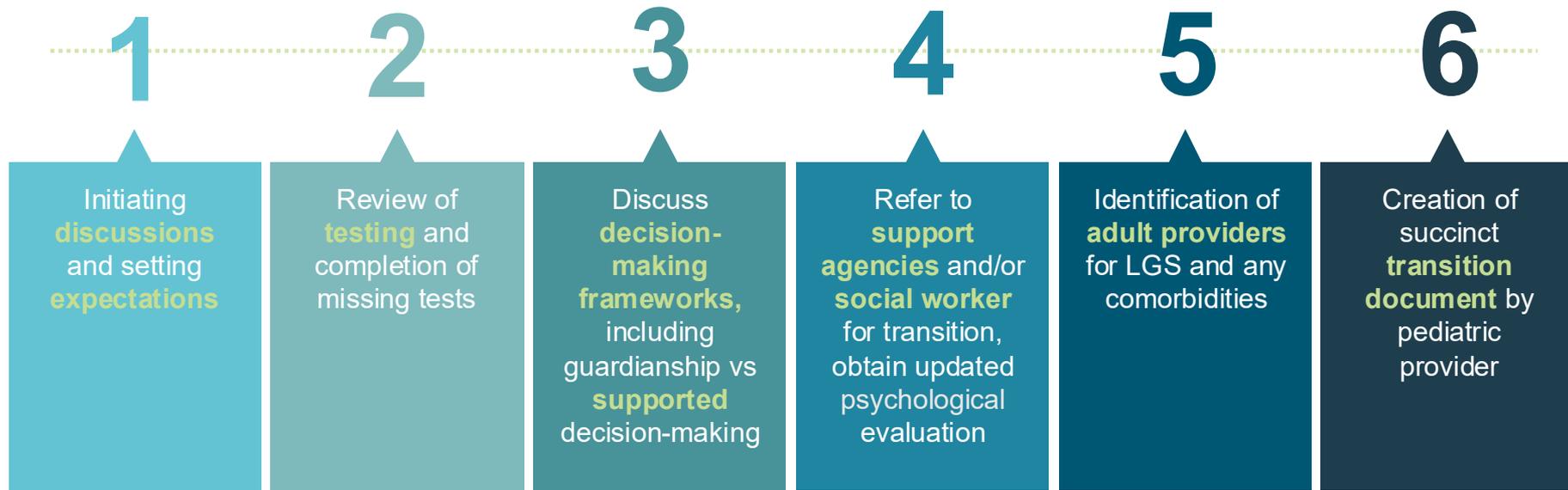
Caregivers are afraid that their aging or health will leave no one able to support their child.

– **Caregiver** of an adult patient with LGS

We tried to transition, but the adult epileptologist didn't know how to manage our child's condition.

– **Caregiver** of a teen patient with LGS

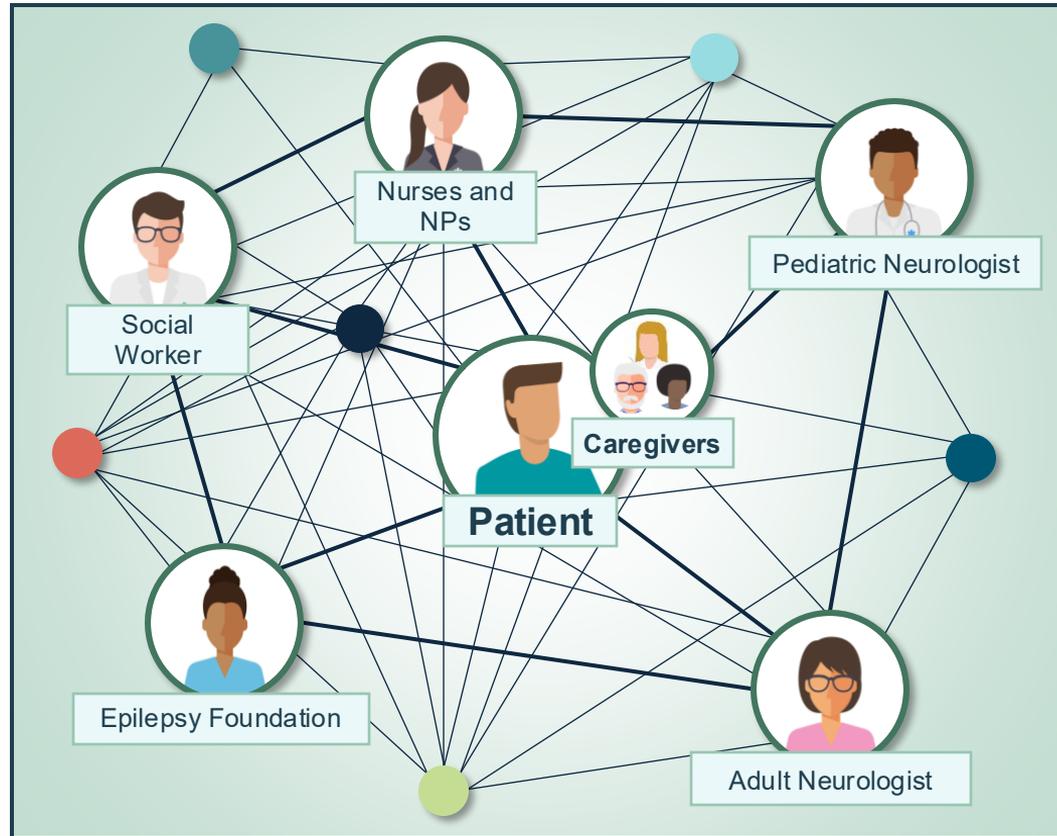
Timeline of Transition Planning



“Group homes were not as effective at taking care of LGS individuals’ needs as the family caregiver would be.”

– **Caregiver** of an adult patient

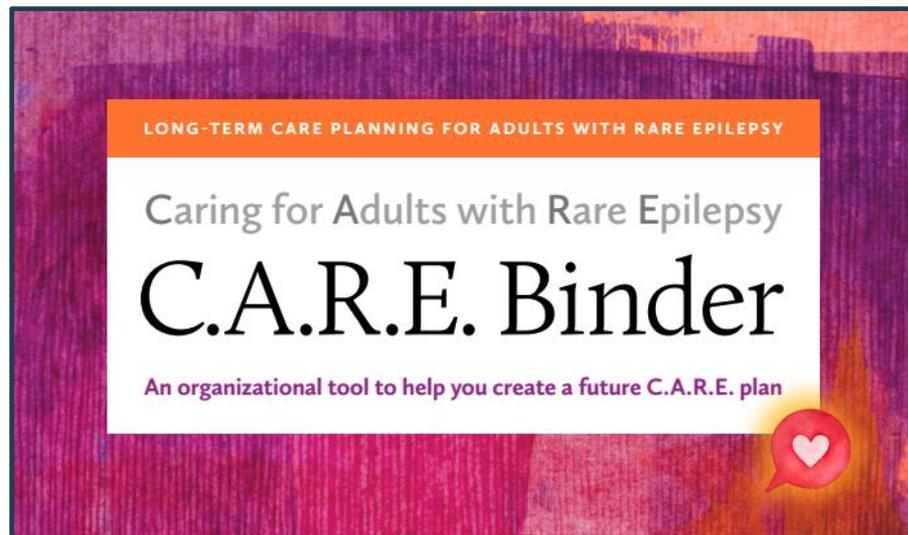
The Multidisciplinary LGS Transition Team



NP = nurse practitioner.

Resource for Transition: C.A.R.E. Binder

- The C.A.R.E. Guide is a resource designed to help families in planning long-term care for LGS
- Includes:
 - Essential factors to consider during the transition process
 - Necessary preparations for adult care
 - Available support services and resources



[https://www.lgsfoundation.org/
adult-care-binder/](https://www.lgsfoundation.org/adult-care-binder/)



Ensuring a Smooth Transition

Faculty Panel Discussion



SMART Goals

Specific, Measurable, Attainable, Relevant, Timely

Put information into action! Consider the following goals, then *set a time frame* that fits with your work environment and *a reasonable improvement target* that aligns with your patient population.

- Monitor patients and caregivers for quality-of-life challenges and address these challenges with resources and support when possible
- Incorporate seizures, non-seizure symptoms, and quality of life into the treatment decision-making process
- Prepare patients and caregivers for transition from pediatric to adult care by initiating discussions early and following key milestones for successful transfer of care



Visit the
Lennox-Gastaut Syndrome Hub

Free resources and education
for health care professionals and patients

<https://www.cmeoutfitters.com/impacting-care-and-quality-of-life-lgs/>

To Receive Credit

To receive CME/CE credit for this activity, participants must complete the post-test and evaluation online.

Participants will be able to download and print their certificate immediately upon completion.

