

A knowledge-to-practice roadmap for measuring quality of life in children with a Developmental Epileptic Encephalopathy

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DEE-P Combined Brain – Workshop 1

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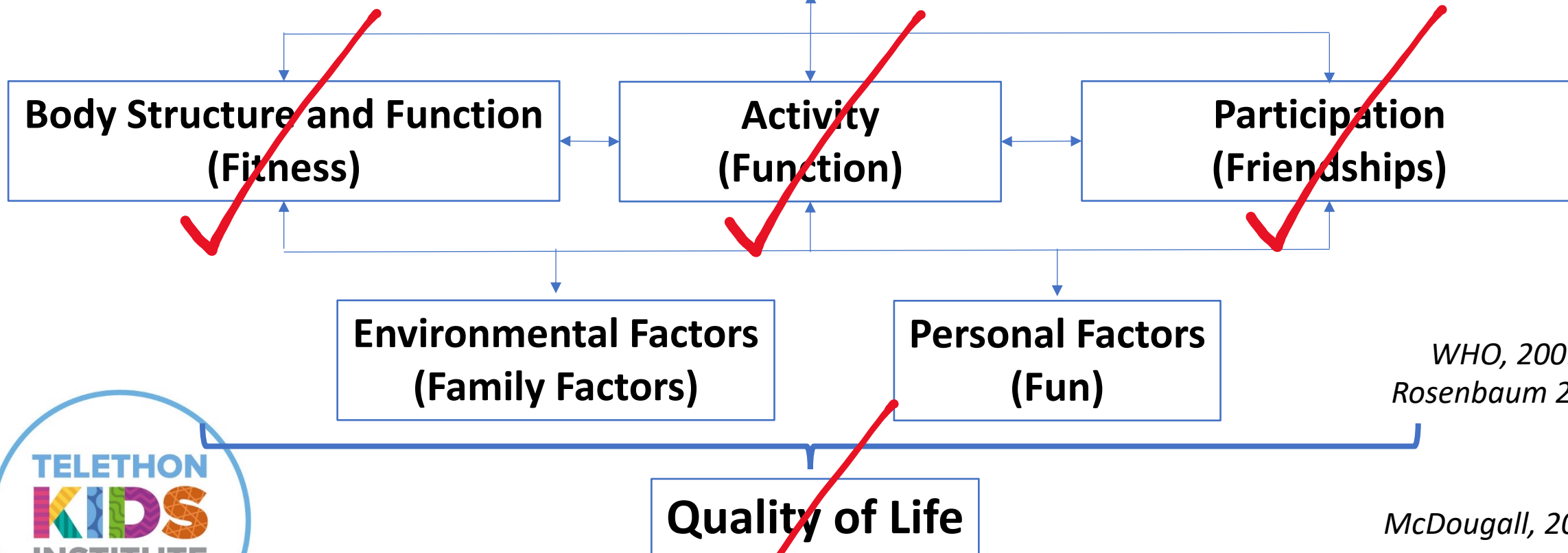
DEEs – needs for reduced symptoms and living well

- Difficulties with
 - Epilepsy
 - Other comorbidities
 - Functional abilities
- **Build on existing strengths** with greater capacity for
 - Learning new skills
 - Enjoying activities
 - Establishing and maintaining social relationships
- **Quality of life** is the feeling of satisfaction with life experiences
 - Is the ideal outcome measure to evaluate treatments and supports

What determines quality of life?

The evidence so far

Children and adults with ID



WHO, 2001
Rosenbaum 2012

McDougall, 2010

The Grand Outcome – Quality of Life

Quality of Life

Affected by
Health
Psychological state
Social relationships
Salient features of their
environment *WHO 2002*

Health-Related Quality of Life

- Focuses on specific impacts of functional status and health on QOL
- Opportunity for more sensitivity
 - Overlap with severity scales *Karimi 2016*
 - Less focus on engagement socially and in the environment

QI-Disability

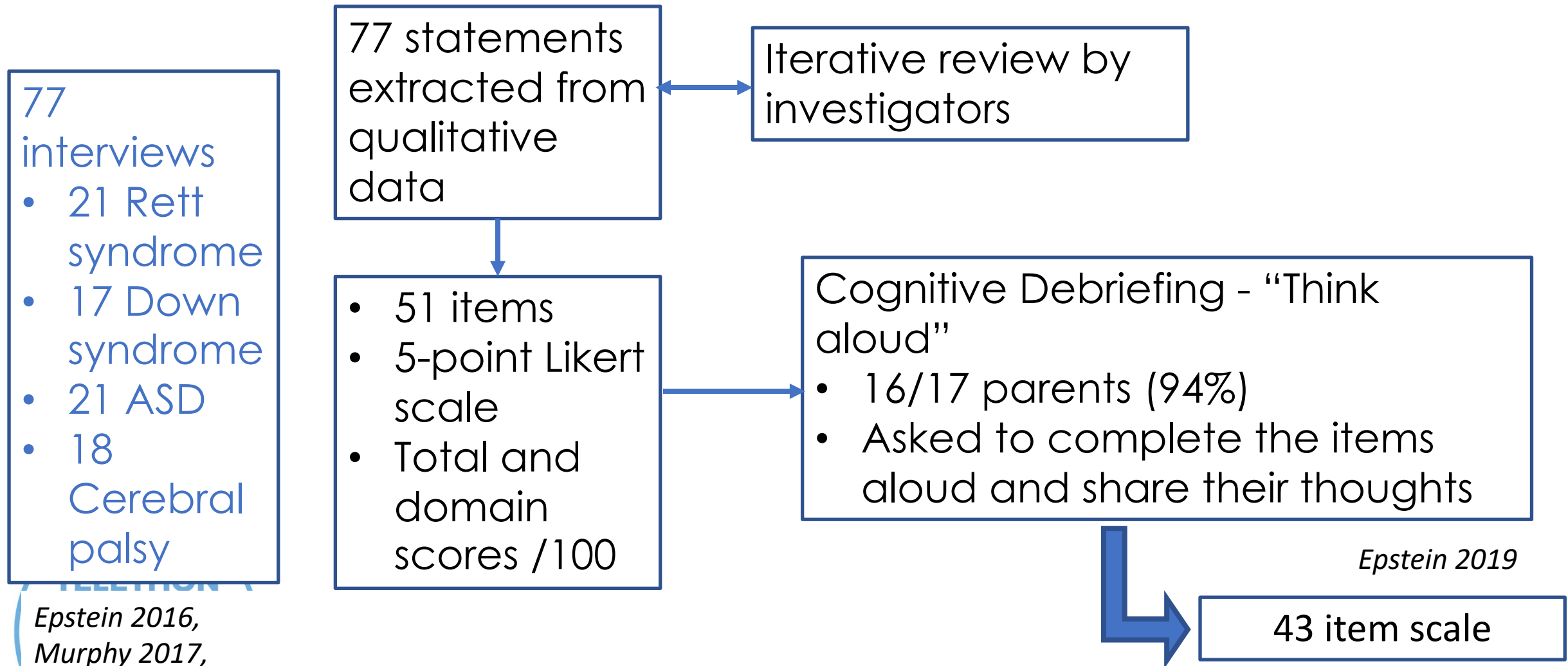
No available HRQOL scale is
sensitive to DEEs

1. To present briefly the development and validation of QI-Disability



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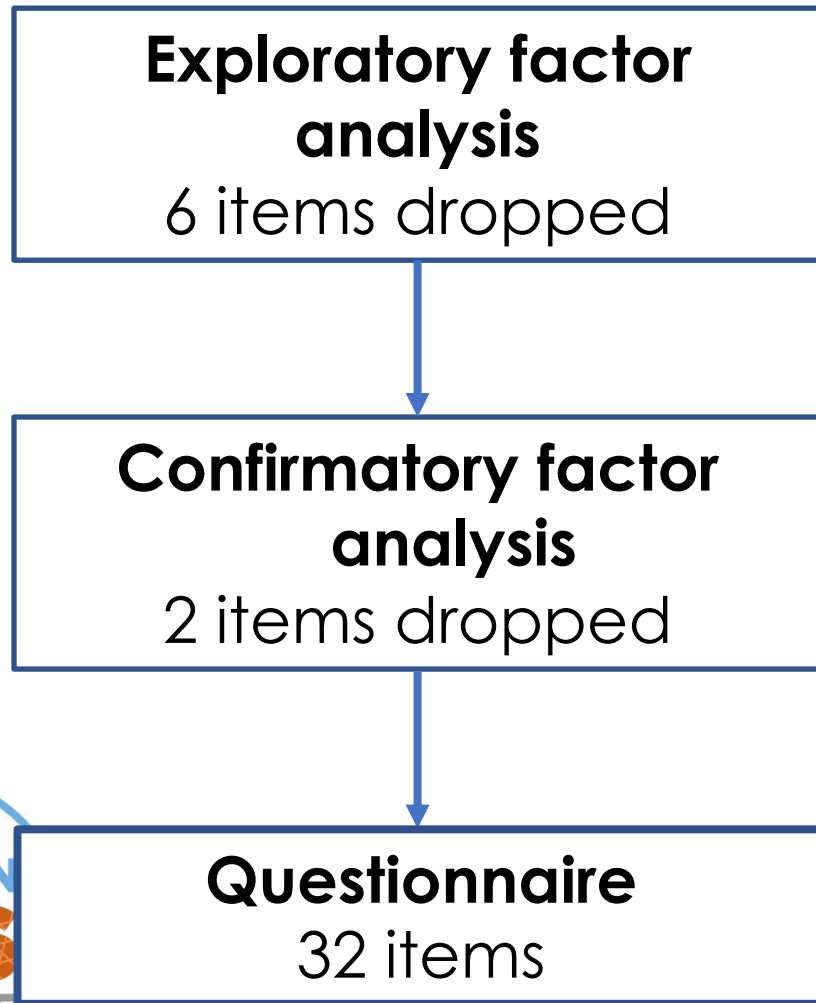
From qualitative data into a scale



Epstein 2016,
Murphy 2017,
Davis 2017,
Epstein 2019

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Pilot study with larger sampling framework (n=253)



- Six-factor structure
- *Factor structure held when the sample split*
 - *Independent walking*
 - *Ability to talk*
- Goodness of fit and discriminant validity - good
- Convergent validity – mostly good

What does QI-Disability look like?

Family and friends

Health and well-being

Over the past month, how often has your child...

1. Had enough energy to participate in d routines and activities
2. Kept in good general health (e.g. avoided coughs, colds, fever)
3. Slept well during the night
4. Been alert and aware during the day

Over the past month, how often has your child...

	Never	Rarely	Sometimes	Often	Very often
16. Expressed happiness when they were understood	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
17. Appeared relaxed when making eye contact	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
18. Initiated greetings with people verbally or nonverbally (e.g. eye contact)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
19. Enjoyed being included	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
20. Enjoyed the social experiences of meal times	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
21. Responded positively when others paid attention to them (e.g. your child smiled, showed interest)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
22. Showed pleasure or excitement when looking forward to activities (e.g. going to school, outings, events)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>



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QI-Disability ready for evaluation tasks for DEEs

Content validity

Items based on family stories
Wording checked with consumers
Domains map to ICF framework

Reliability and responsiveness

Responsive to changes in physical health and behaviour
Good stability on retesting

Minimal detectable difference

A change of 5 points on a 100-point scale identifies change

Validity

Total and domain scores map to expected differences

- diagnosis
- functioning
- comorbidities

Vision

Every individual with a DEE will live with strong QOL

QI-Disability

To support evaluations

2. To consider available HRQOL tools for DEEs and present preliminary data

Collaborators in the US

- Prof. Avani Modi
- Dr. Brandy Fureman
- Prof. Jeffrey Buchhalter



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HRQOL measures for DEEs

Epilepsy-related HRQOL measures *Crudgington 2020*

- Systematic review – leading candidate measures (not for IDD)
 - Quality of Life in Childhood Epilepsy (QoLCE)
 - Health-Related Quality of Life Measure for Children with Epilepsy (CHEQoL)

Relevance to IDDs

- Revision of ELDQoL *Buck 2007*
- PedsQL Epilepsy module – limited for intellectual disability

Follensbee-Junger 2016, Modi 2017

Scope of domains

- Common domains
 - Seizure severity
 - Side effects
 - Neurological function
 - Mood and behavior
 - Cognition
 - Executive functioning
- Less common domains
 - Social interaction
 - Activities
 - Choice and control (independence)

Scope of items

- e.g., PedsQL Epilepsy module
- Impact domain
 - *Hard to do daily tasks on own as other kids*
- Cognitive domain
 - *Hard to understand what I read*
- Executive function
 - *Hard to stay organized*

Gap for DEEs

First qualitative study

Modified grounded theory

- 24 parent caregivers of
- 22 children,
 - 2 to 18 years
 - 12 females
 - 9 with CDD, Dravet syndrome or Rett syndrome
 - All had daily seizures, 12 >5 per day
 - Range of comorbidities and functional abilities
- 6 domains



Data collection

Rushil Shah

Physical Wellbeing

- Activity and energy
- Fatigue and sleep
- Play

She'll help me lift her arm into her T-shirt. A lot more helpful and she is more aware of her surroundings of what's going on. When she's out for the count, pretty much she's like, dead weight. – 6-year-old girl

Behavior

- Emotional regulation
- Expressing emotions

We got this activated toy for Christmas where she hits a button, and she's laying there and she hits it. And her eyebrows just went right up right when it starts dancing. Her communication of her happiness is from her cute little eyebrows– 7-year-old girl

Social engagement

- Family interactions
- Social connectedness

*“There's jokes on the eye-gaze device. She loves telling **"Knock-knock" jokes**. ... **She loves making the kids laugh** and she gets the teachers laughing too.” – 11-year-old girl*

*If she's feeling good. She loves when people talk to her. Yeah, that's probably one of her favourite things, as well as just having people talk to her and tell their stories and stuff. **She's just like, lights up when they talk to her**— 2-year-old girl*

Enjoyment of activities

- Music TV games
- Nature and animals
- Outdoor activities

*We just got back from visiting my parents. They have a house in the mountain. So we go there a lot and she does enjoy that. She does really well. **We did a little Santa Sleigh Ride. And she was awake the whole time. She was chatting, like her little cooing sounds and yeah, she really enjoyed that.** – 18-year-old girl*

Learning

- Experiential learning
- Opportunities for learning and engagement

*we had a good month... **he was kind of learning to grasp things again, and kind of bringing his cup up to his mouth again. And then the seizure started back and he lost it again.** – 5-year-old boy*

Independence

- Making own choices
- Self-sufficiency

*School will say she's a different kid. **She's just awake, she will get on her Toby - the eye gaze - she says things that she needs to say, you know, it's very minor, but big for us. You know, she can make choices –***
18-year-old girl

*He's more independent. **He's more willing to give things a go and he can entertain himself. But when he's struggling, it's no, I can't get anything done. I have to actually be with him. Yes, he might just want to watch his iPad and do not much.**– 8-year-old boy*

The case for an epilepsy specific HRQOL measure

- Imperative for strong outcome measures for children with DEEs, including QOL and HRQOL
- Epilepsy-related HRQOL scales
 - Not grounded in grass roots experiences for DEEs
 - Are not well scaled

QI-Disability for QOL

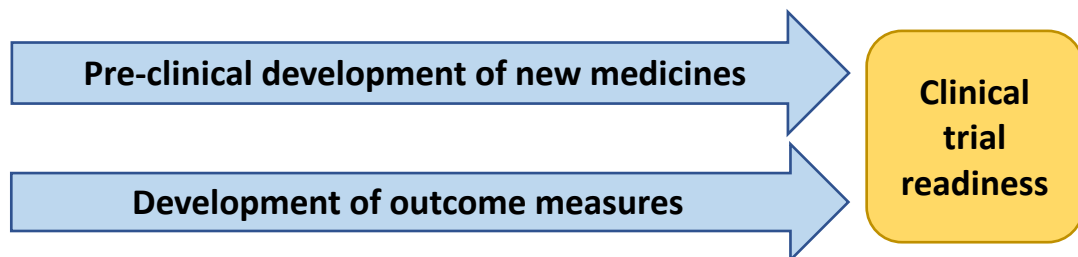


QI-Epilepsy for HRQOL - sensitive to DEEs

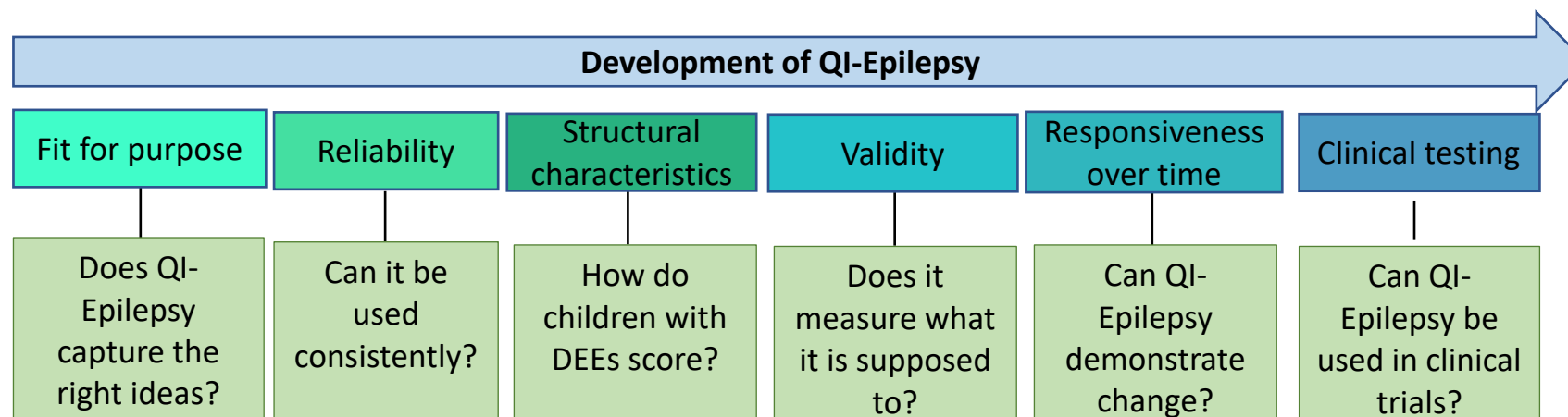


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How to create an excellent HRQOL measure for DEEs



Processes



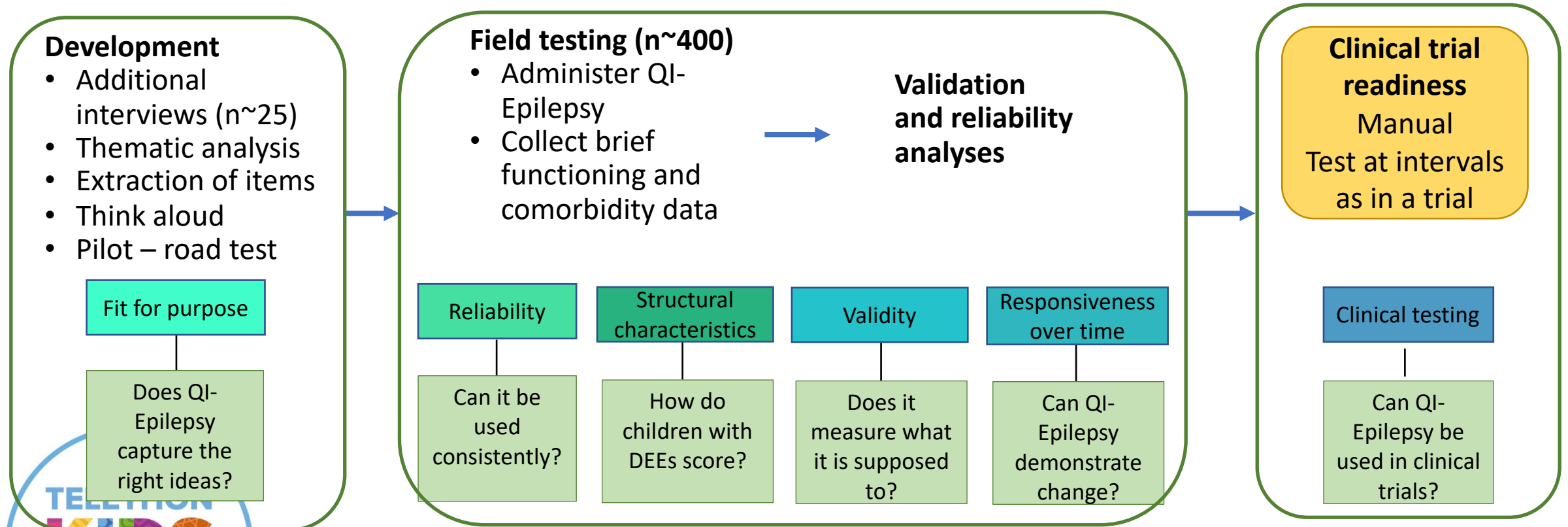
Actions



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Necessary tasks

Collaborators in the US: Prof. Avani Modi, Dr. Brandy Fureman, Prof. Jeffrey Buchhalter



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