

# Nurse-Led Mental Health Monitoring in 2-5 Year-Old Children with CF Initiating elexacaftor/ tezacaftor/ ivacaftor (ETI) at a Single Canadian CF Center

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# Background

- Mental Health (MH) side effects including *sleep disturbances, concentration problems, and worsening psychological functioning* are reported in adolescents and adults who initiate ETI<sup>1,2</sup>
- Less is known about possible MH side effects in the pediatric population

***“Negative side effects including sleep disruption and worsening psychological function should be **monitored and evaluated systematically** in people with CF who initiate treatment with ETI.”***

<sup>1</sup> Bathgate CJ, Muther E, Georgiopoulos AM, Smith B, Tillman L, Graziano S, Verkleij M, Lomas P and Quittner A (2023). Positive and negative impacts of elexacaftor/ tezacaftor/ ivacaftor: Healthcare providers' observations across US centers. *Pediatr Pulmonol* 58: 2469-2477.

<sup>2</sup> Graziano S, Boldrini F, Romana Pellicano G, Milo F, Majo F, Cristiani L, Montemitro E, Alghisi F, Bells S, Cutrera R, Fiocchi AG, Quittner A and Tabarini P (2024). Longitudinal Effects of Elexacaftor/ Tezacaftor/ Ivacaftor. Multidimensional Assessment of Neuropsychological Side Effects and Physical and Mental Health Outcomes in Adolescents and Adults. *Chest* Vol 165 (4): 800-808.



January 2024:

ETI approved for 2-5 year old  
pwCF in British Columbia



How will you know?

Will it impact my child's mental health?

Could side effects appear when they're older?

How will it impact my child's mental health?

## ***Initiating ETI in the Young Child***

“Too Young!”




“Too well!”

“Too New!”



# CF Mental Health Services

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- 
- 
- Community psychiatrist
  - Community Clinical Counsellor
  - CF Mental Health Nurse Navigator
  - CF Social Worker
  - CF Nurse Clinician
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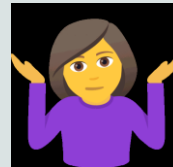


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- + Psychologist!
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*CF Pharmacist  
Extraordinary Eva!*





- Literature / Eva
- Dr. Quittner
- CF Social Worker

Mental Health Screen (“Emotional Wellness Screen”)

Mental Health Symptoms

# Pediatric Quality of Life Inventory (PedsQL)

## Parent Report for Toddlers Ages 2-4

In the past **ONE month**, how much of a **problem** has your child had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in active play or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Bathing	0	1	2	3	4
6. Helping to pick up his or her toys	0	1	2	3	4
7. Having hurts or aches	0	1	2	3	4
8. Low energy level	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad or blue	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Playing with other children	0	1	2	3	4
2. Other kids not wanting to play with him or her	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

Scoring:

>75% = Normal

64-75% = Mild

50-63%=Moderate

<50% = Severe

Preschool  
Pediatric  
Symptom  
Checklist  
(PPSC)

PRESCHOOL PEDIATRIC SYMPTOM CHECKLIST					
Recommended 18 month, 0 days to 65 months, 31 days		Your Name:			
		Relationship to the child:			
These questions are about your child's behavior. Think about what you would expect of other children the same age, and tell us how much each statement applies to your child.					
		Please mark under the heading that best fits you			
			Not at all	Somewhat	Very much
Does your child...	Seem nervous or afraid?	0	1	2	
	Seem sad or unhappy?	0	1	2	
	Get upset if things are not done in a certain way?	0	1	2	
	Have a hard time with change?	0	1	2	
	Have trouble playing with other children?	0	1	2	
	Break things on purpose?	0	1	2	
	Fight with other children?	0	1	2	
	Have a hard time calming down?	0	1	2	
	Have trouble staying with one activity?	0	1	2	
Is your child...	Aggressive?	0	1	2	
	Fidgety or unable to sit still?	0	1	2	
	Angry?	0	1	2	
Is it hard to...	Take your child out in public?	0	1	2	
	Comfort your child?	0	1	2	
	Know what your child needs?	0	1	2	
	Keep your child on a schedule or routine?	0	1	2	
	Get your child to obey you?	0	1	2	
	Total Sum (across Columns)				
Do you currently receive counseling or behavioral health services? If yes, where?				Yes	No
Are you interested in talking with someone about your child's behavior or development?				Yes	No
For Office Use Only					
To Score					
1. Score the items		"Not at all" = 0, "Somewhat" = 1, "Very much" = 2			
2. Find the summed total		Any summed score of 9 or more means that families might like to talk about how to learn more about their young child.			

Scoring:

“Any score of 9 or more means that families may wish to talk about how to learn more about their young child”.



## ETI MH Monitoring 2-5 Yrs

**Emotional Wellness Screen:**  
PedsQL

**CFQ-R**  
Emotion domain

**Mental Health Symptom Tracker:**  
Preschool PSC

Preschool Anxiety Scale, if needed\*

\*If scores high on  
PedsQL Emotional  
Functioning

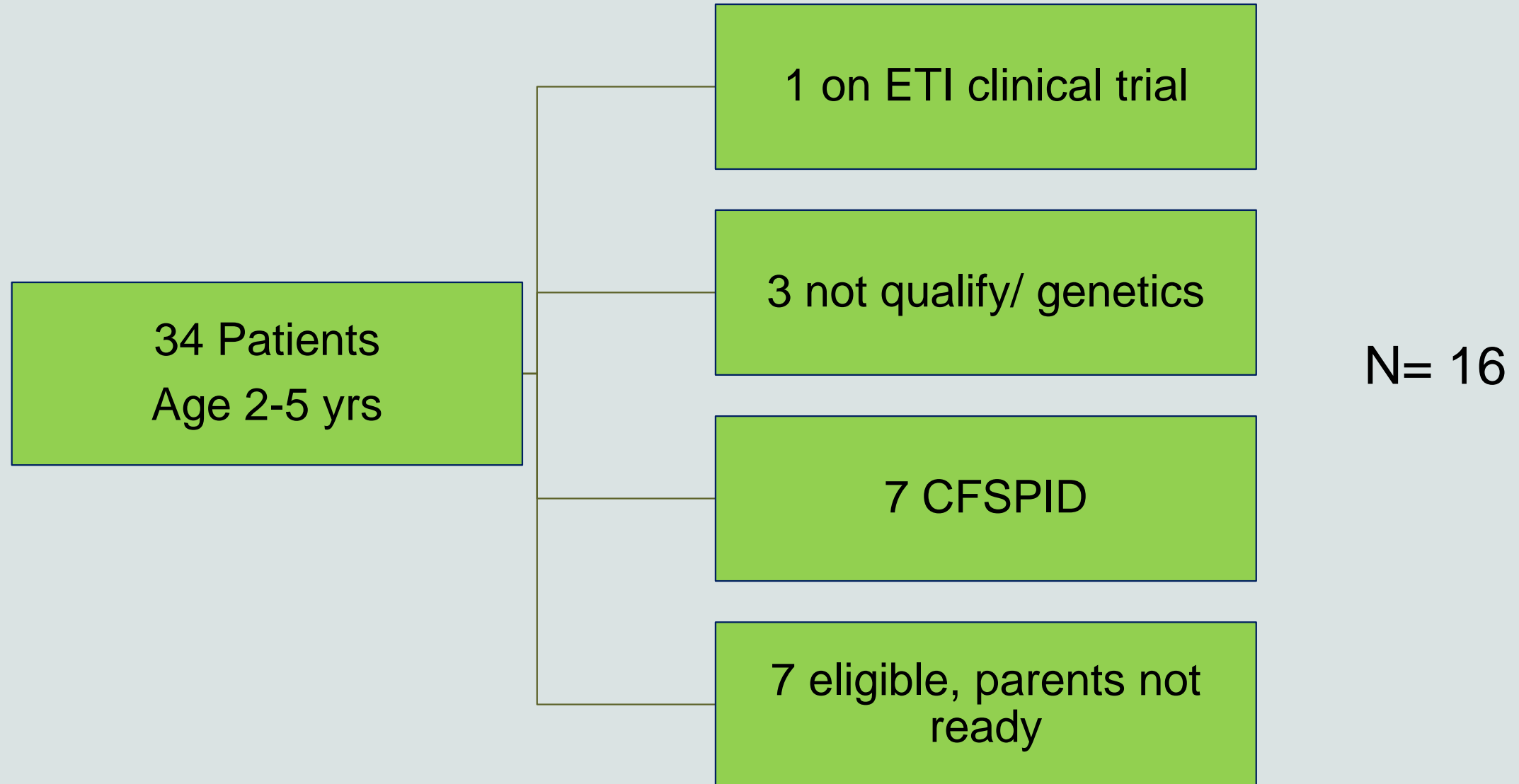
\*Do at:

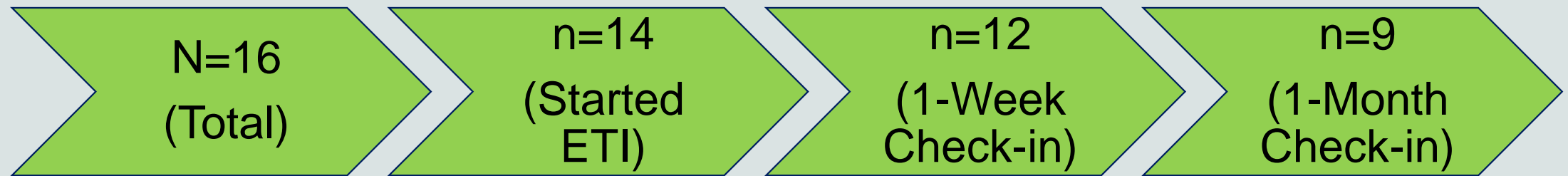
- Baseline
- 1-Month
- 6-Month
- 12 Month

Telephone Check-in:

- 1-week
- 1-month

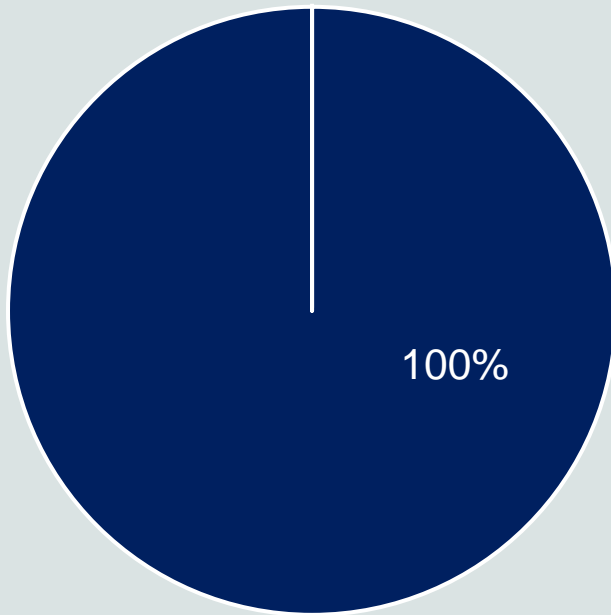






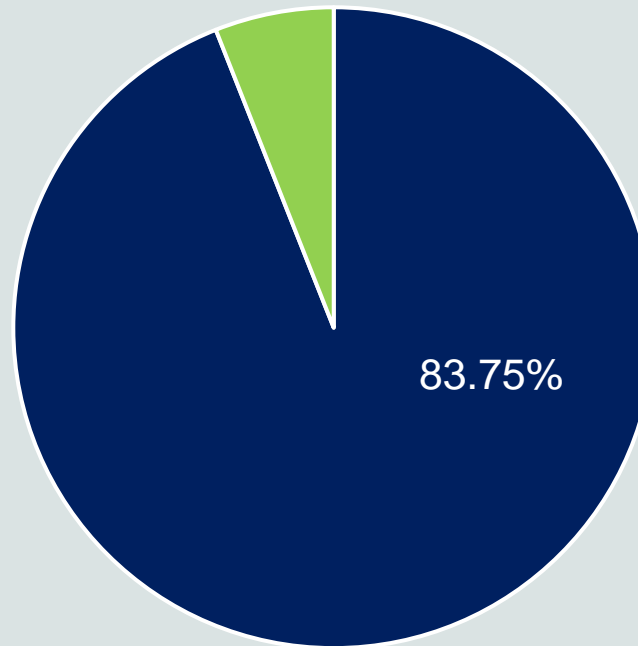
# Results: Baseline (N=16)

CFQ-R Completed



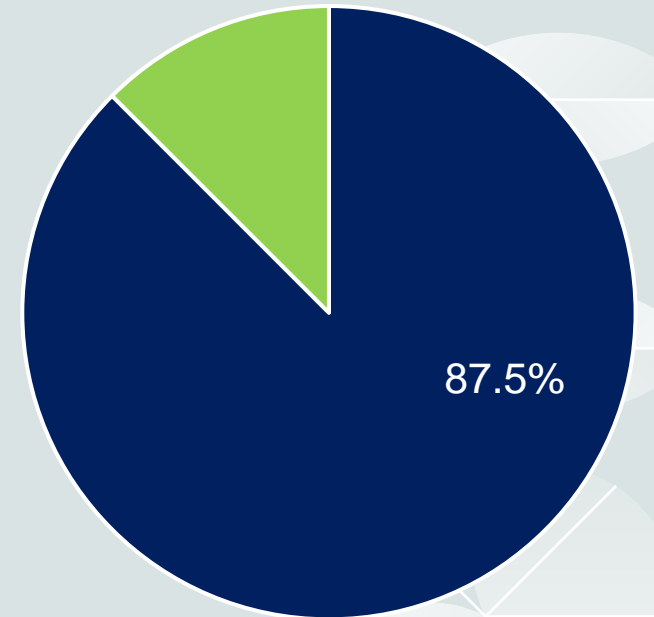
■ Completed ■ Not Done

PedsQL Completed



■ Completed ■ Not Done

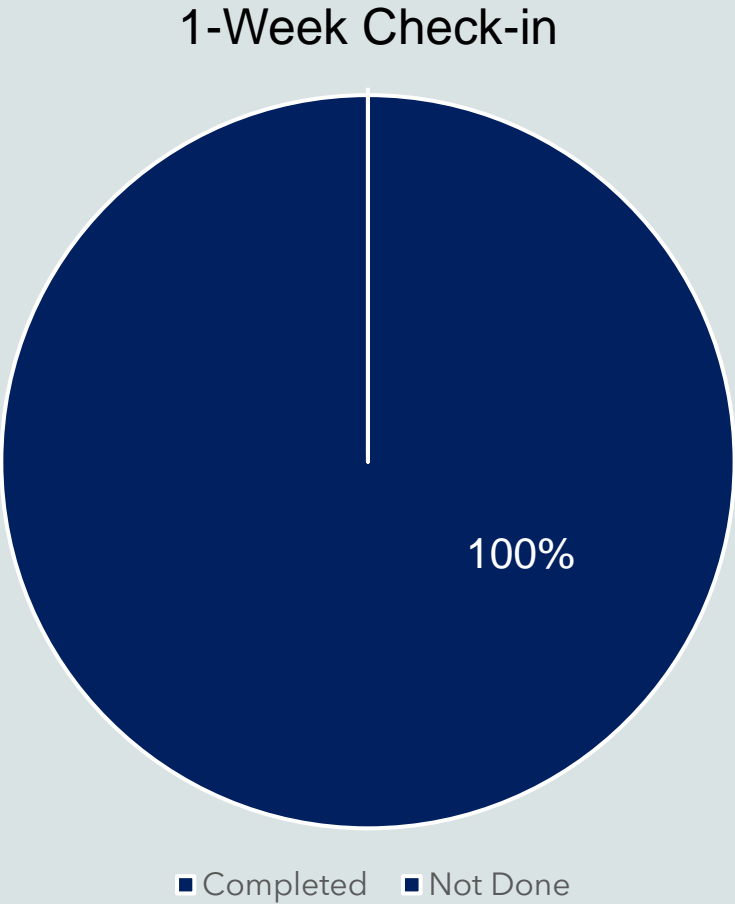
PPSC Completed



■ Completed ■ Not Done

# Results: 1-Week Phone Check-in (n=12)

- Sleep?
- Emotions?
- Concentration?
- Hyperactivity?
- Other?



# Less likely side effects which might happen:

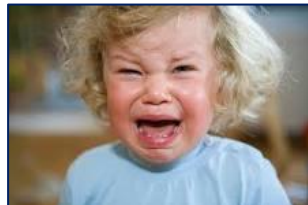
- Headache



- Feeling dizzy



- “Big emotions”



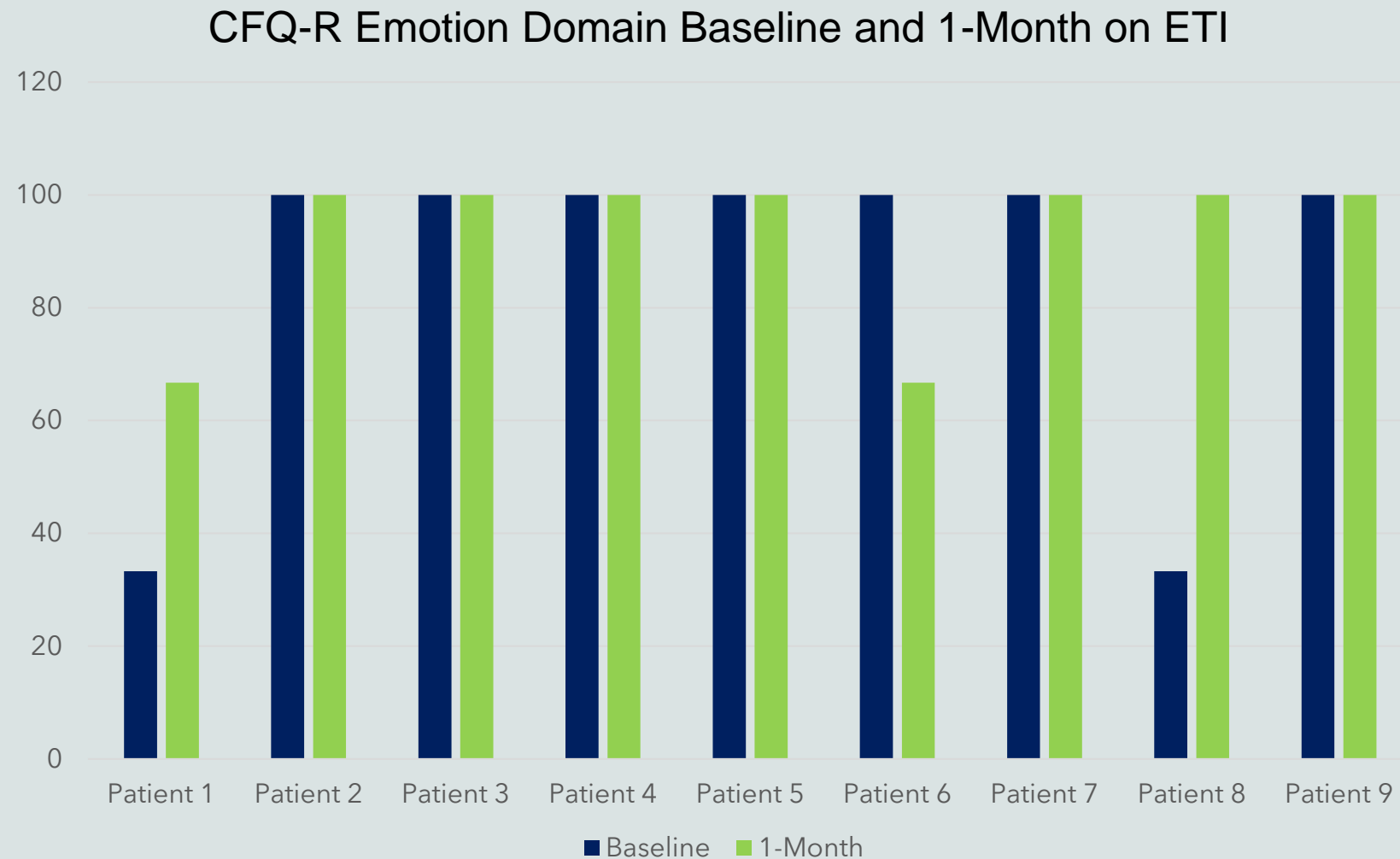
- Feeling hyper



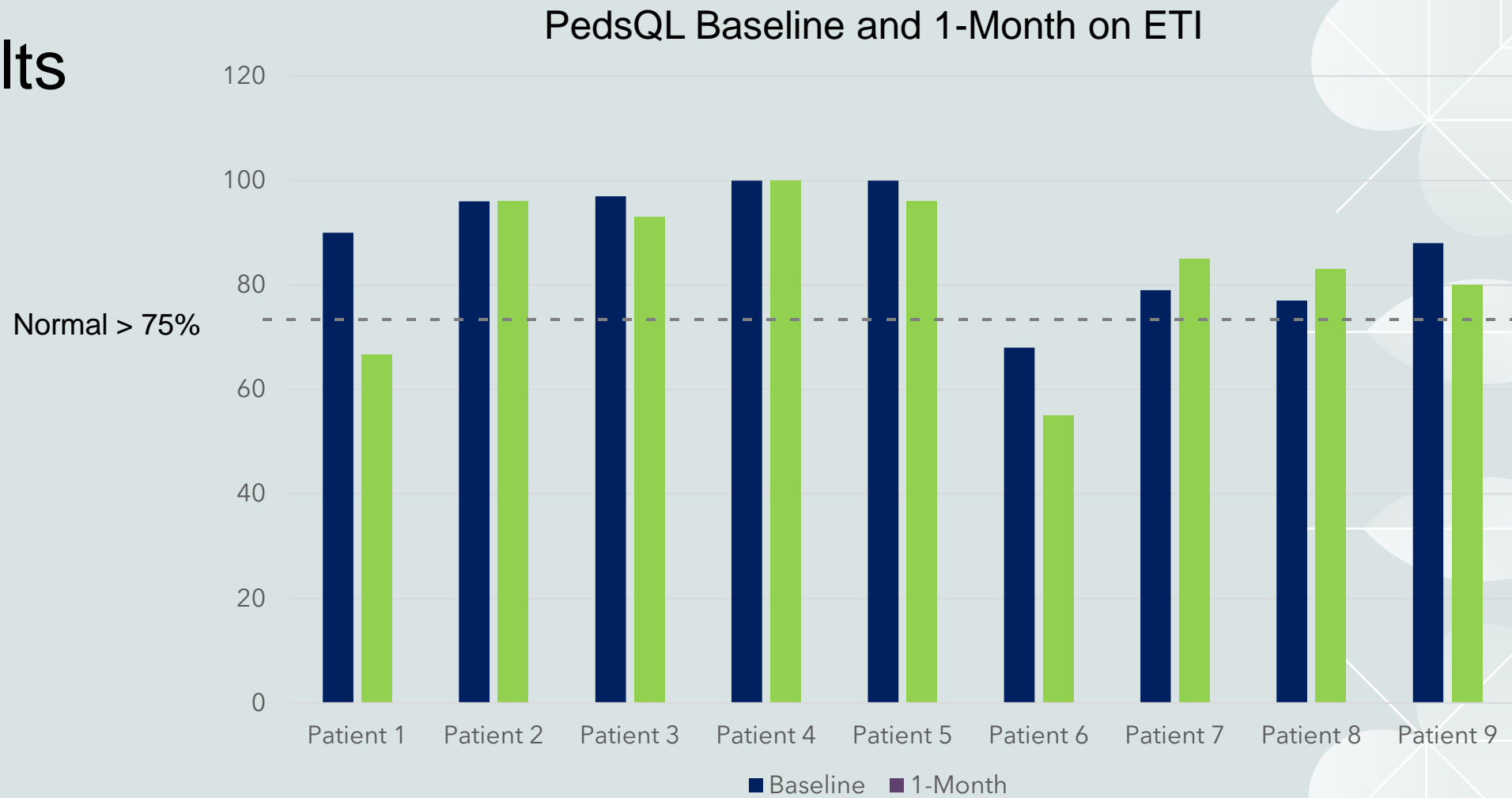
- Troubles with sleep



# Results



# Results



# Results

## Baseline:

- URTI
- Transition from crib to bed

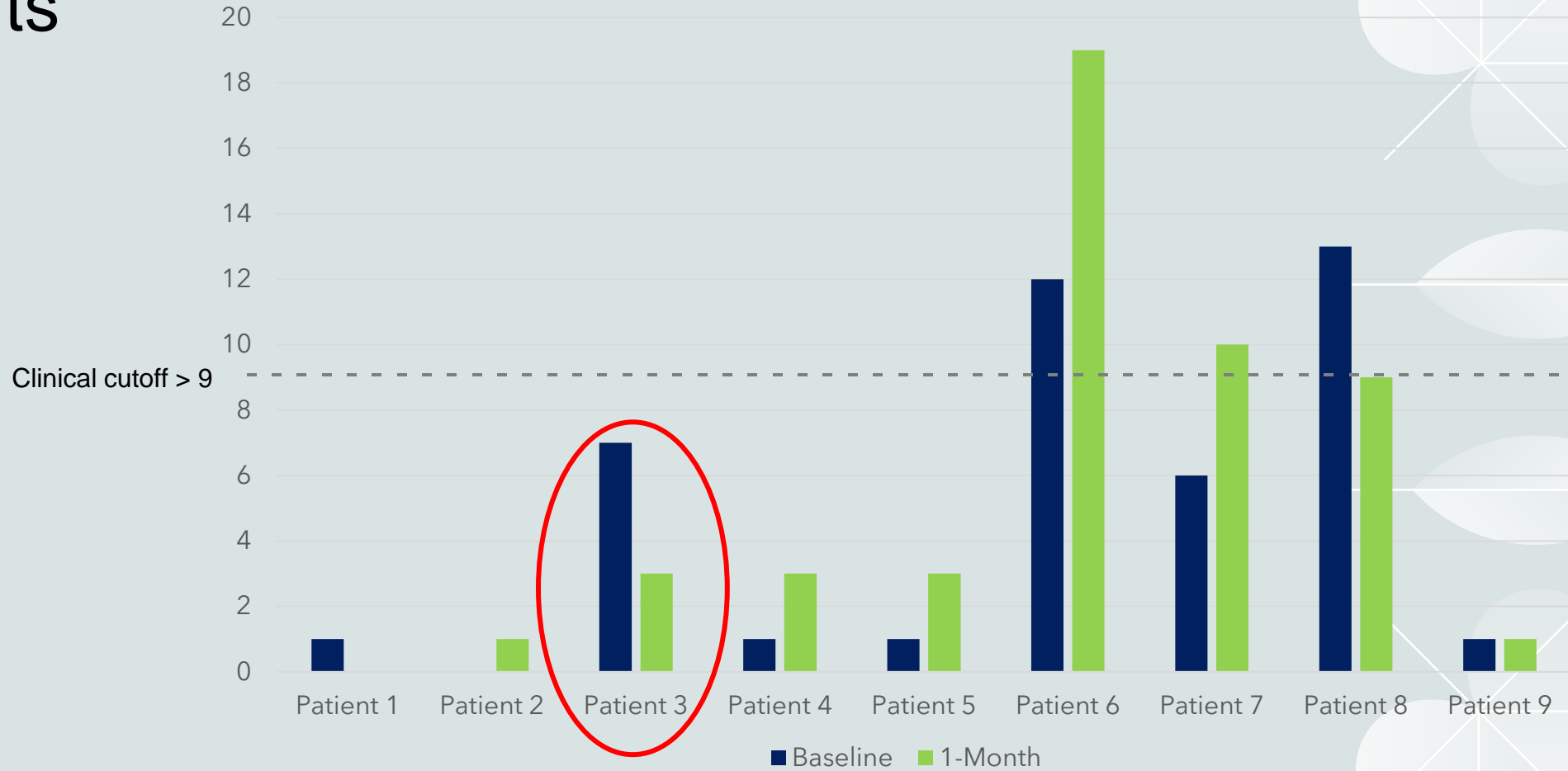
## 1-Month:

- New tics

## Action:

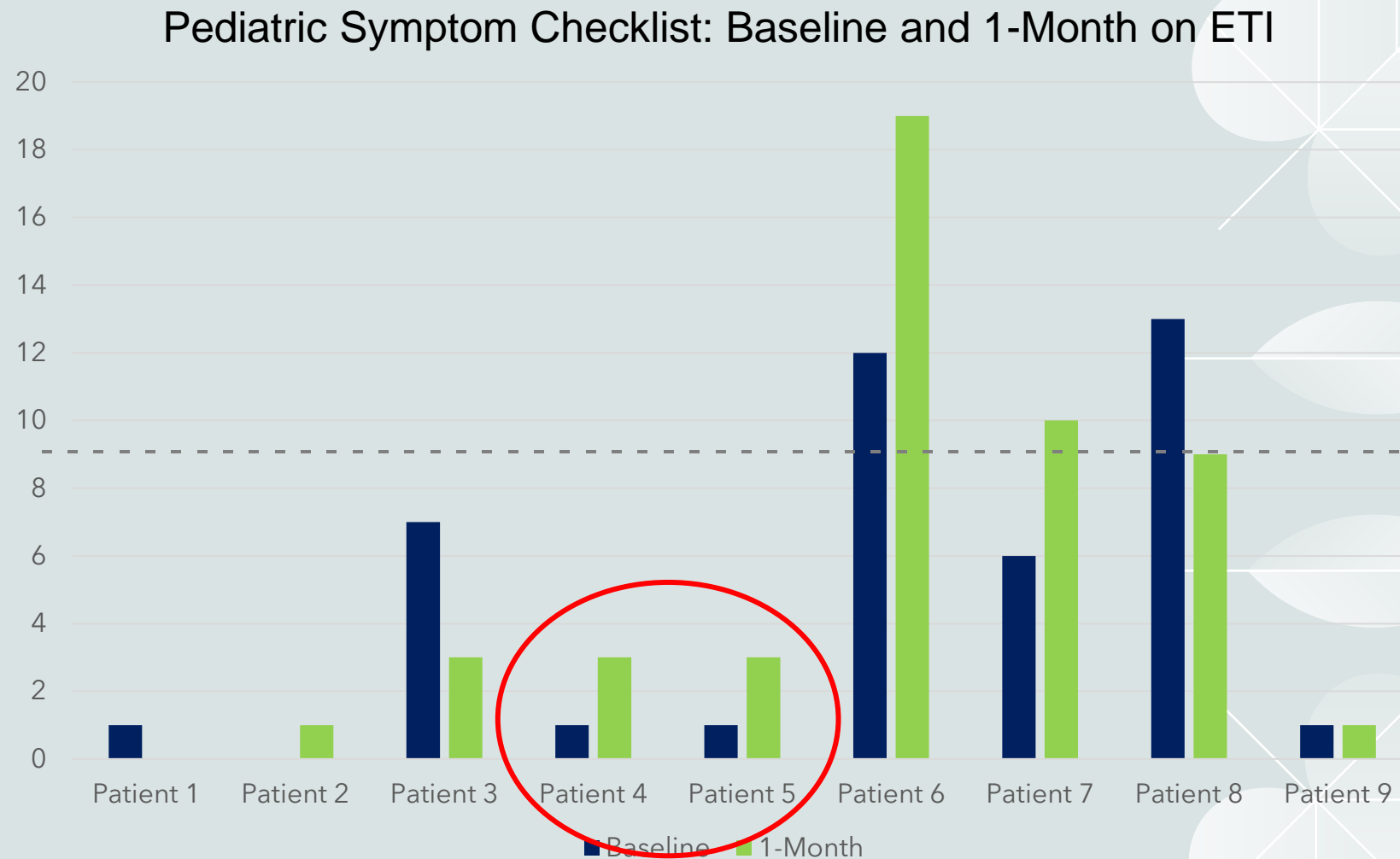
- "Watch"

Pediatric Symptom Checklist: Baseline and 1-Month on ETI





# Results



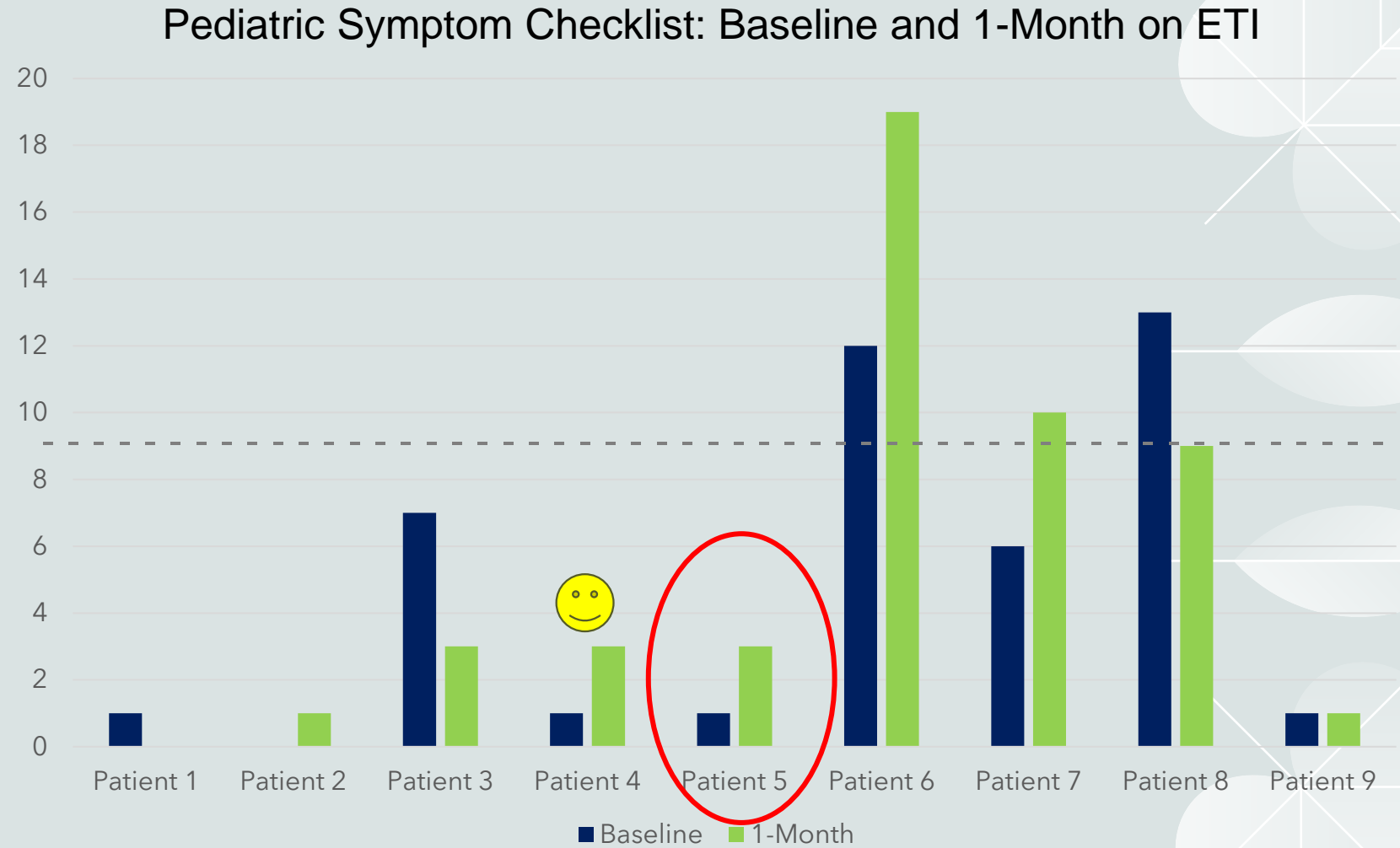
# Results

## 1-Month:

- Anger outbursts
- Sleep Disturbances

## Action:

- Flip AM/PM doses



# Results

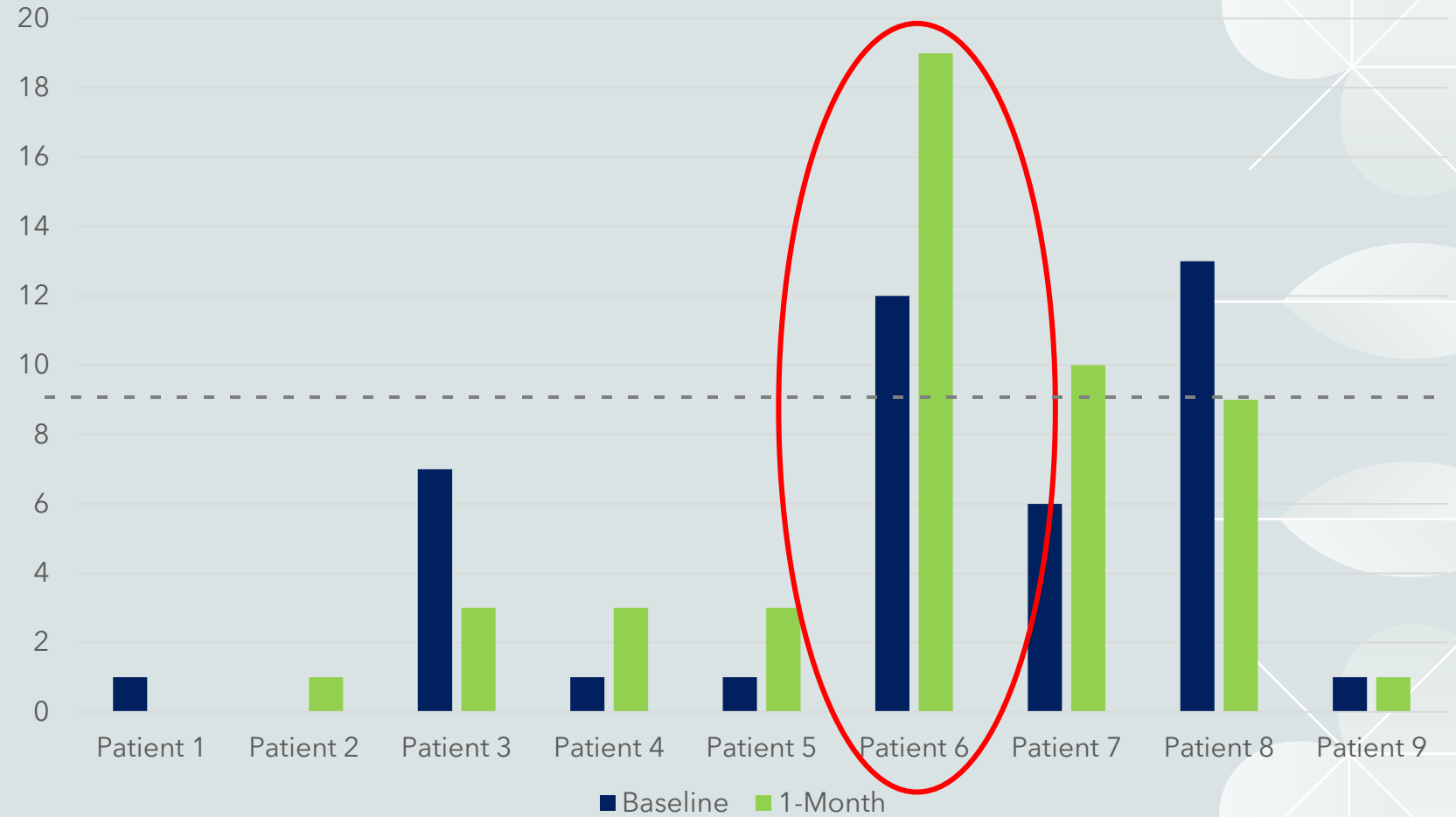
Pediatric Symptom Checklist: Baseline and 1-Month on ETI

1-Month:

- Anger outbursts

Action:

- Flip AM/PM doses



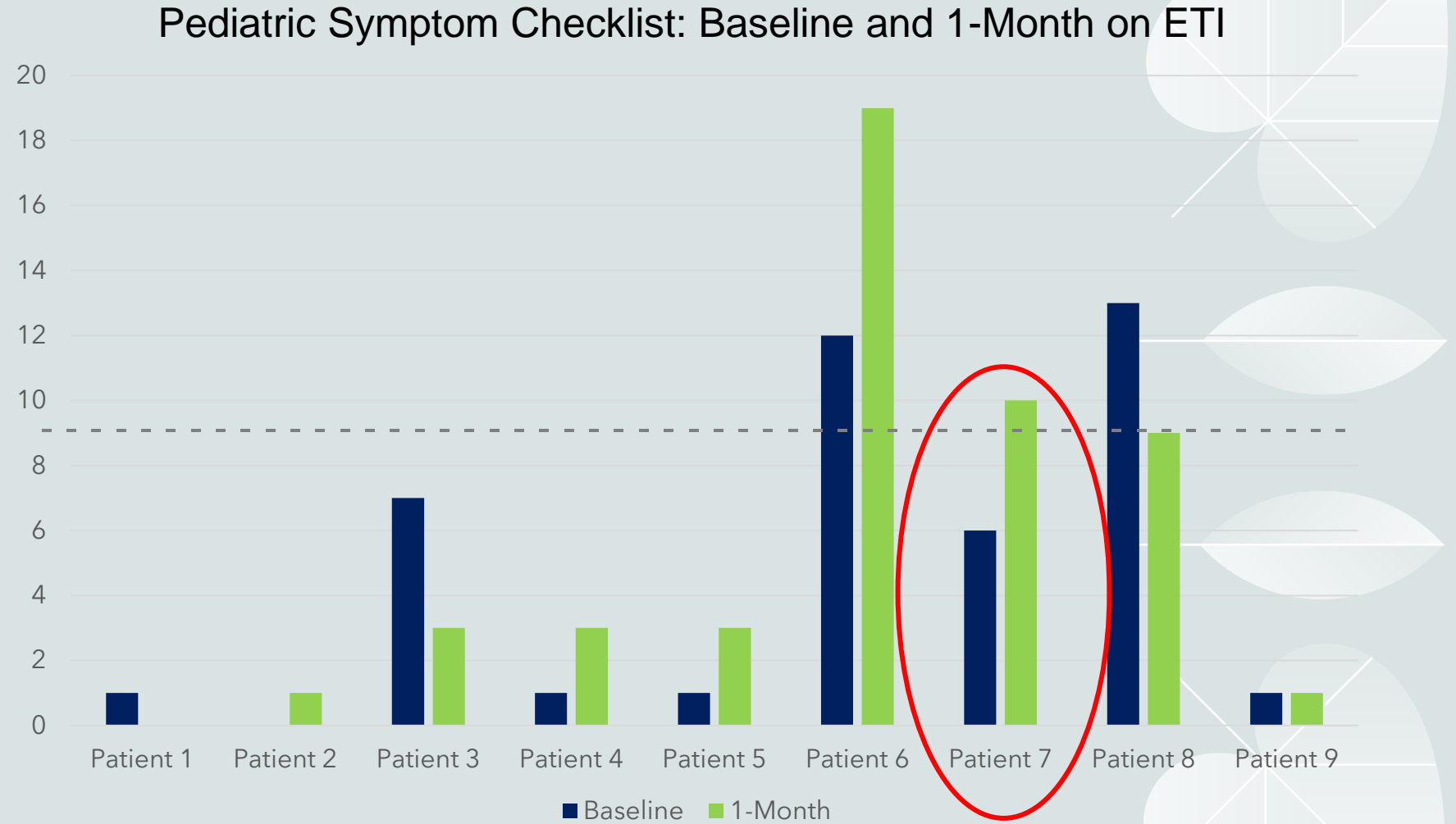
# Results

## 1-Week:

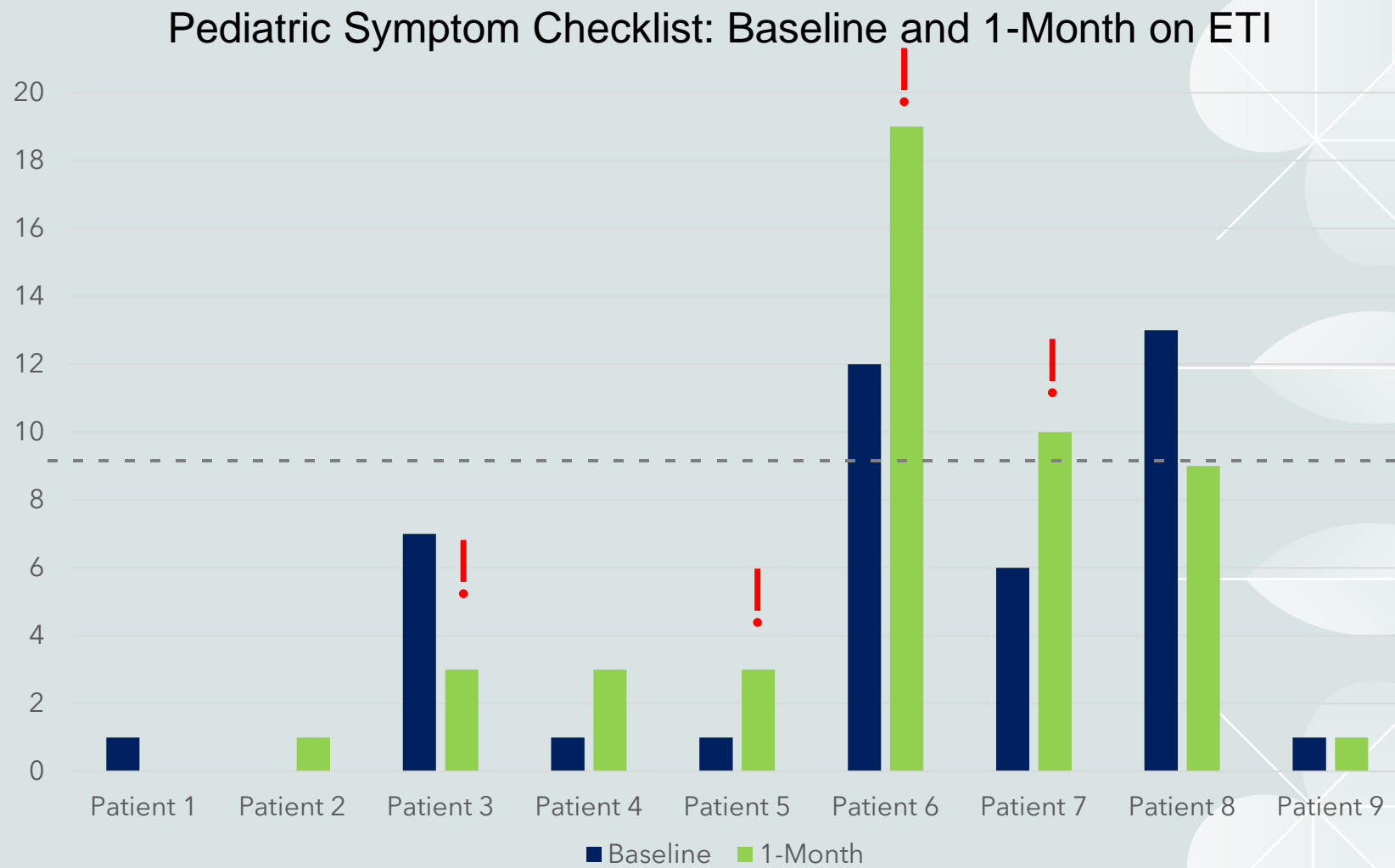
- Anger outbursts
- Tactile issues

## 1-Month:

- “Resolved”

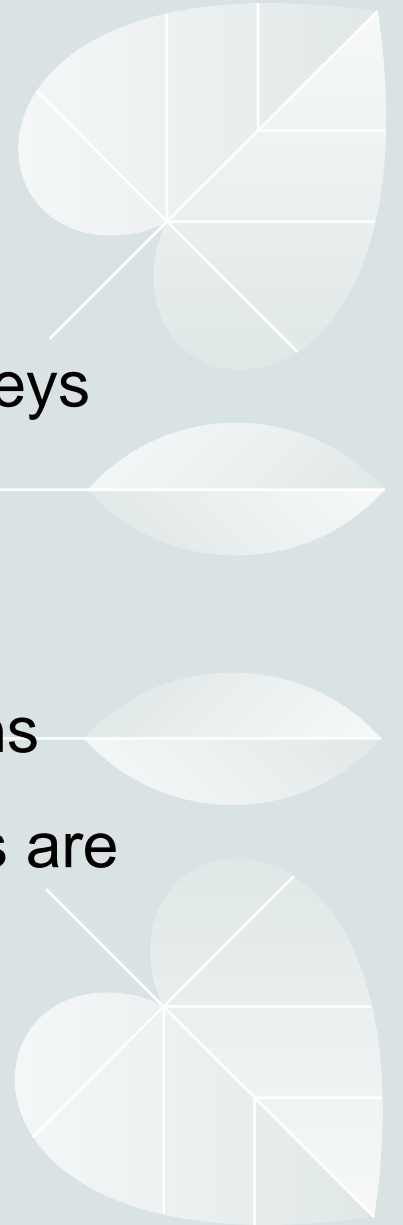


# Results



# Preliminary Conclusions

- Good job of capturing baseline/ 1- week/ 1- month data
- Patients who require interpreter services are missing surveys at a higher rate
- Too many questionnaires!
- No protocol (yet) for reducing/ stopping ETI for MH reasons
- What to do when questionnaires don't match what parents are telling us?



# Preliminary Conclusions

- MH side effects started 2-10 days after ETI initiation
- Probable ETI MH side effects:
  - ❑ **sleep disturbances**
  - ❑ **extreme anger outbursts**
  - ❑ **tactile sensitivities**
- Possible ETI MH side effect: **new eye tics**
- Questionnaires may miss ETI-related MH side effects
- To date, flipping AM and PM doses improved symptoms in 2 (out of 2) children



# Preliminary Conclusions

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➤ Team approach needed:

- Parent/ Caregiver
- Pharmacist
- Nurse
- Physician/ NP
- SW
- Psychologist







# Takeaway Points

We don't have all the answers yet...but we're working on them



## Reassurance:

- ❖ Parents know their children best
- ❖ They will be listened to
- ❖ We have strategies to try

Monitor for side effects before and after ETI starts:

- ❖ That are atypical for that child
- ❖ That occur within a stable life context

Monitor for side effects after dose increases?

