

CERTAINTY OF GOALS: WHEN THE OUTCOME IS UNCERTAIN FOR CHILDREN WITH SEVERE NEUROLOGICAL IMPAIRMENT

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Objectives

- Discuss problems due to severe impairment of the central nervous system that can remain intractable to treatment options
- Define tipping points that can prompt anticipatory discussions, including suggested recommendations that best meet goals
- Utilize language strategies that assist families when prognosis is not possible



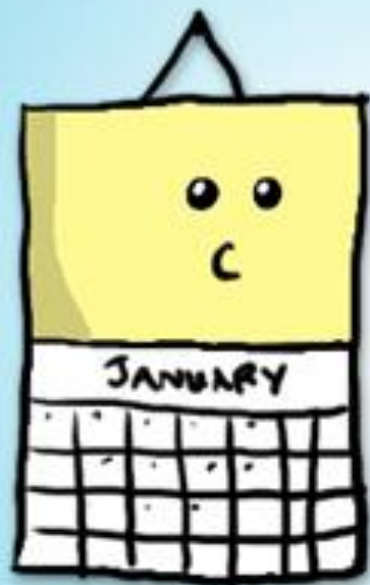
Courageous
Parents
Network:
videos with
his mother

Challenges with Decision Making

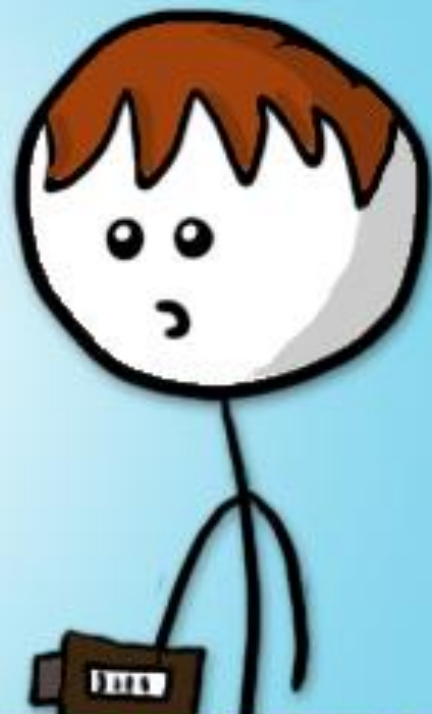
- Prognosis: prediction of probable course
- Wide range of outcomes
- 2 survival phases at birth or injury
- No biomarkers
- **Goal: lessen decisional regret**

Leong K 2013, Merker B 2008, Whiteford 1996

GIVE IT TO ME
STRAIGHT, DOCTOR



YOU'VE GOT
TWELVE MONTHS



Prognosis

Palliative Care

Outcome based (death, development) and **Decision based** (surgery, DNI/DNR)



Process based (define goals, review options, attend to emotions, allow hope)

Uncertainty to Certainty

Uncertainty of Outcome: life span, recovery, benefit from intervention



Certainty of Decision: purpose, intent, and goal of a decision

Hope

- **Adaptive:** to cope with adversity and loss
- **Affirmative:** to convey commitment and love

Feudtner 2009, Feudtner et al 2010



I've learned from
my son that
everything is
possible, it doesn't
mean everything
will happen.

Quality of Life (QOL) Considerations

- QOL as indicated by parents of 14 children with profound intellectual and motor disability (PIMD)
 - Ability to enjoy
 - Comfort
 - Awareness
 - Multiple health problems
 - Falling below the lower threshold of QOL

Zaal-Schuller IH, et al. Considering quality of life in end-of-life decisions for severely disabled children. *Res Dev Disabil.* 2018;73:67-75.

End of Life Decision Making (EOLD)

- **Parents:** half indicated missed opportunity by physicians to discuss EOLD at time of stability
- **Physicians:** found it difficult to identify when a parent was “ready” for discussion
- **Physician discussion:** typically initiated at time of acute deterioration

Zaal-Schuller IH, et al. How parents and physicians experience end-of-life decision-making for children with profound intellectual and multiple disabilities. Res Dev Disabil. 2016;59:283-293.

Patterns of End of Life Care

- **Progressive** disorders of the CNS *versus* **Static** encephalopathy, congenital, genetic
 - **Static** group less likely to have palliative care consult, die at home, plan location of death, or have DNR orders in place
 - **Static** more likely to receive intensive life sustaining therapies in last 2 days of life

34 deaths: general characteristics

- Average age: 22 years
- Age range:
 - ▣ 11 months to 50 years
 - ▣ 10 (29%) older than 30 years
- Conditions: anoxic and traumatic brain injury, neuro-structural malformations, in-utero infection, genetic, metabolic

General Characteristics

Feature / Problem	N=34
Feeding tubes	N=34 (100%)
Anti-seizure drugs	N=33 (97%)
Complex symptom management	N=23 (68%)
Respiratory: chronic, recurrent	N=15 (44%)
Gastrointestinal: acute recurrent	N=10 (29%)
Ventilation: invasive, noninvasive	N=7 (21%)

Goals Discussion N=27 (79%)

- **Triggers:** symptoms, decreased engagement in activities, health burden, EOL features
- **Triggers added:** functional ileus
- **Common goals:**
 - ▣ To be comfortable, To not suffer
 - ▣ To get better, To live as long as possible
 - ▣ To be with family and loved ones

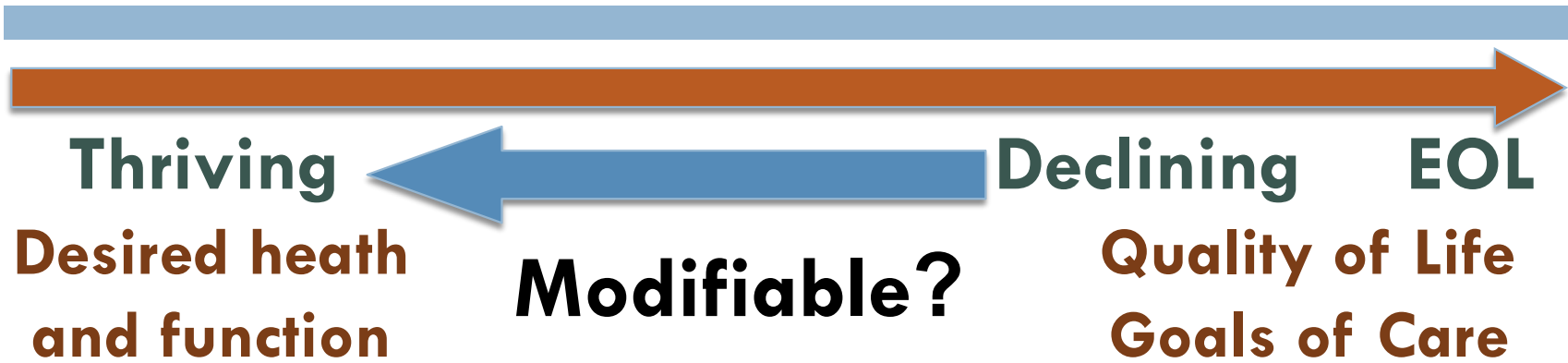
Redirection of Care in 23 of the 27

Process: days, months, years

Time until death: days to 5 years

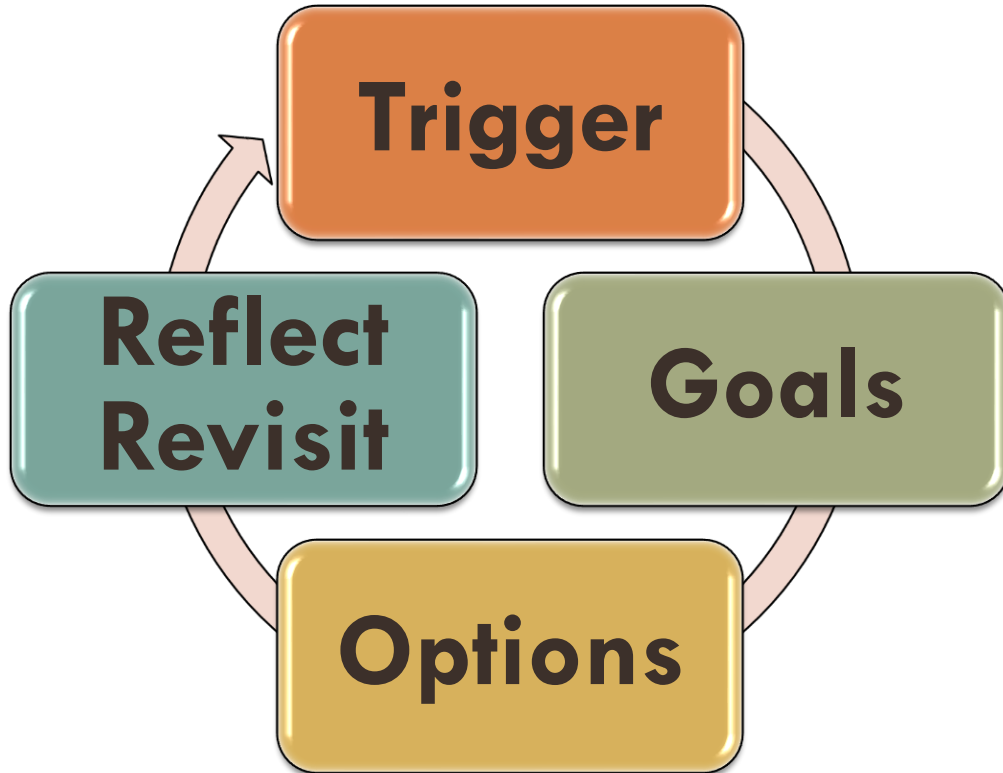
Reasons for Redirection	N= 23 of 34
General decline	N=19 (55%)
Global symptom burden	N=16 (47%)
End of life (EOL) features	N=16 (47%)
Respiratory: chronic, recurrent	N=15 (44%)

Quality of Life and Goals of Care



Natural tension between problems without cure
that we intend to improve though may not
have the hoped for outcome

Iterative Discussion Loop



Triggers for meeting

- Significant changes
 - ▣ Symptoms
 - ▣ Quality of life
 - ▣ Health
- End-of-life features
 - ▣ Edema
 - ▣ Functional ileus

Ileus or Edema at End of Life

- Ileus and/or edema at **End-of-Life**: 13 (38%)
 - Acute or recurrent ileus: N=10 (29%)
 - Edema: N=6 (18%)
- **Assessment**: metabolic panel, urine analysis and culture
 - **11: pre-existing decline in health, function, QOL**
 - 2 with severe anoxic brain injury
- Ileus with **recovery**: stable QOL and health

Siden H, et al. Pediatric enteral feeding intolerance: a new prognosticator for children with life-limiting illness?

Hauer J. Feeding Intolerance in Children with Severe Impairment of the Central Nervous System

Language *and* Pauses to Listen

- “These features worry me...”
- “The tests were normal. It is unlikely that there is a fixable reason for his intestines shutting down. This can be due to the body shutting down or changes in the area of the brain that regulates the gut. In some, gut function won’t return.”

Hauer J. Feeding Intolerance in Children with Severe Impairment of the Central Nervous System: Treatment and Prevention. *Children (Basel)*. 2017; 5(1). pii: E1.

Language *and* Pauses to Listen

- “We will give an amount of fluid that his body is more likely to tolerate. We will discuss what that means if he doesn’t...”
- “I’m glad we are protecting him from feeds/fluids that his body can’t process at this time”
- Decision to hold feeds/fluids: no regret, “lesser of 2 evils”, “the only thing that made any sense”

Neurophysiology is Complex

- Hypothalamus, Thalamus, Midbrain, Pons, Medulla regulation
 - ✓ Heart rate, blood pressure, temperature
 - ✓ Arousal
 - ✓ Vasomotor (hypothalamus, medulla)
 - ✓ Intestinal motility, vomiting
 - ✓ Respiratory center

End-of-Life (EOL) Decision Making

- “I don’t believe in mights.” Does not help to consider “what might happen”
- Mom’s coping: focus on how well he will do
- Revisit: if goal of comfort cannot be met or intervention will not meet physiological goal

Goal Concordant EOL Care

- Need for around the clock nursing
- Requires experts in EOL care
- Active adjustments in care plan
 - ▣ Symptom management
 - ▣ Adjust feeds / fluids at end of life
 - ▣ Stop nonessential interventions

HEALTH / Quality of Life

A Point in Time



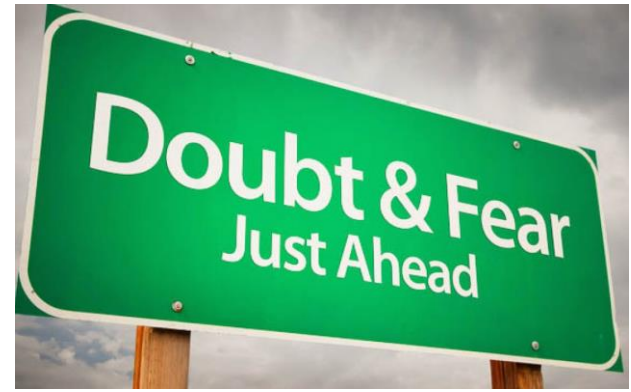
HOPE

2 trajectories at the beginning

- Hours to weeks, occasionally months
- Those who live months often live years

Time to “relearn” their child

TIME = days, weeks, months, years



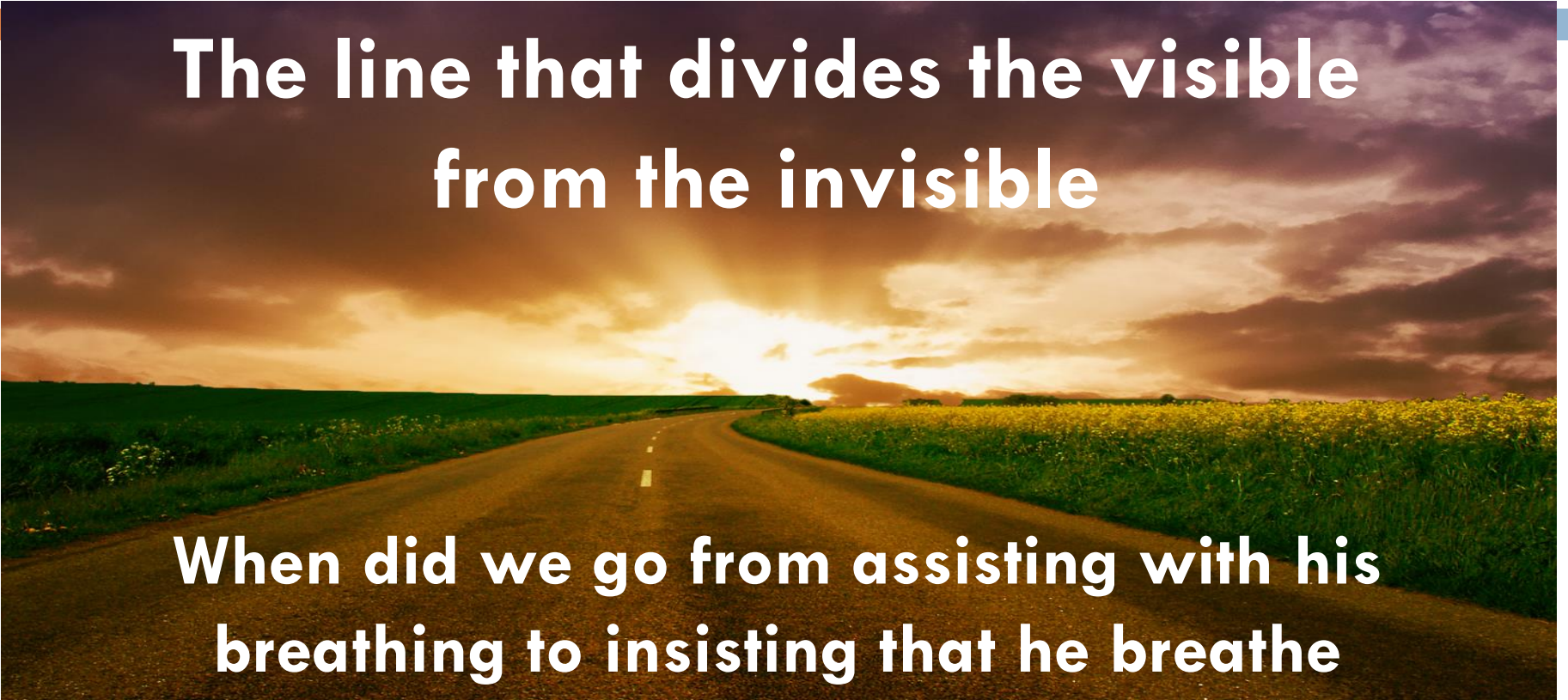
What can we “measure”

- Create an explicit process for triggers
- What goals are intended by the intervention
 - ▣ Life extending *and...*
 - ▣ See what outcome is possible, attempt to modify health and/or quality of life
- Celebrate success!
- Patience, reflect and revisit goals as needed

Goals of Care – the Horizon

**The line that divides the visible
from the invisible**

**When did we go from assisting with his
breathing to insisting that he breathe**



Conclusions and considerations

- Define triggers: monitor for changes in quality of life and burden of treatment
- Determine Goals of Care, revisit as needed
- Offer goal concordant treatment plans
- End-of-life care requires expertise and intermittent around the clock nursing

Thank you!

Keep in touch!

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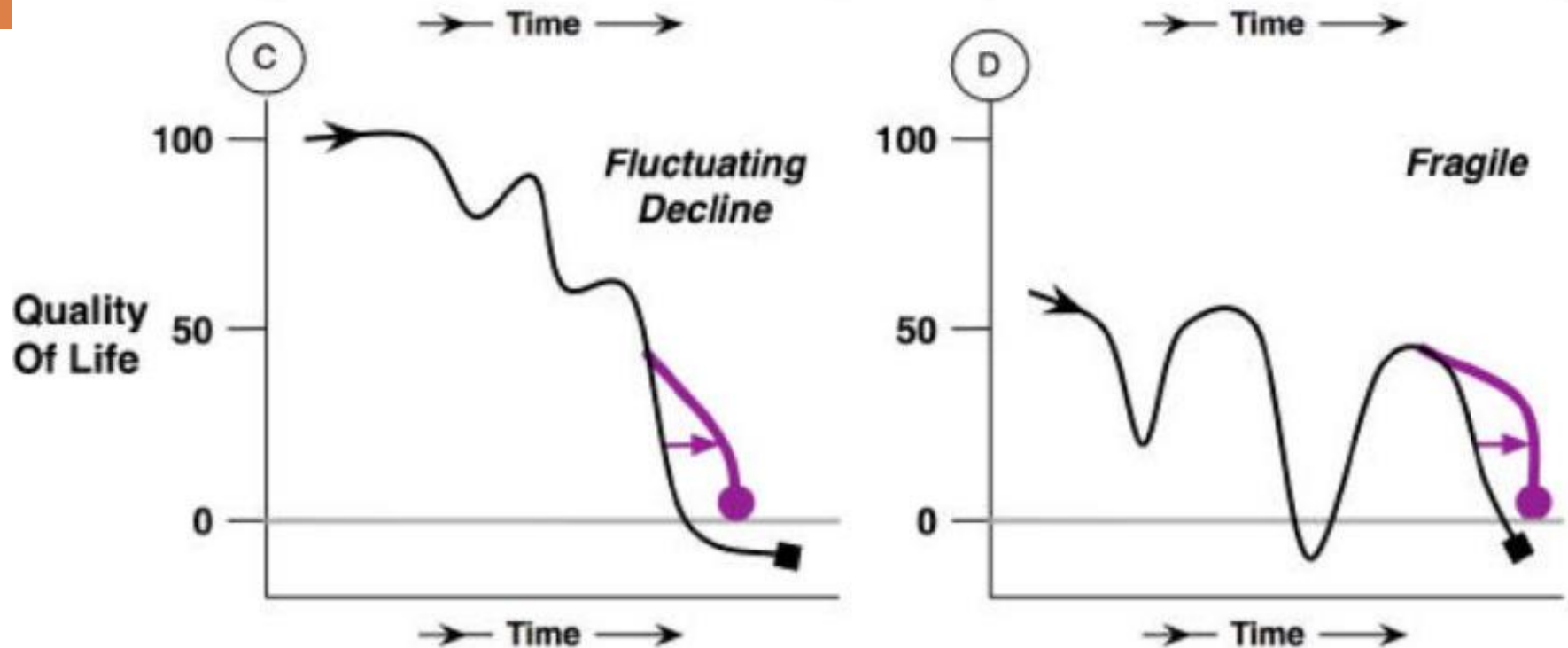
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- An Official ATS/AACN/ACCP/ESICM/SCCM Policy Statement: Responding to Requests for Potentially Inappropriate Treatments in Intensive Care Units: <https://www.thoracic.org/statements/resources/cc/inapprpr-ther-st.pdf>

Quality of Life (QOL) Considerations



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Respiratory Features

- Respiratory distress: active respiratory effort and unmet symptom burden, uncommon at EOL
- Opioid use in 15:
 - ▣ 13 (86%) on scheduled morphine: months to years
 - ▣ “I think he lived longer because of this”
- Acute and chronic treatment
- Change in respiratory rate and rhythm at EOL:
 - ▣ apneic pauses, Cheyne-Stokes, agonal

Goals of Care and Decision Making

- Altered QOL (alertness, comfort)
- Hopes/Goals: improved health, alertness, and comfort
- “I’ve heard you mention several hopes, let’s review what might be possible”

Goals of Care and Decision Making

- Follow-up: “I wish...” “I wonder...”
- Discussion when technology not offering the benefit it once did
- “I’m glad for all the years of benefit...”
- Introduced with no need for decision

Language Suggestions

- **Acknowledge the emotion:** I see your distress, I am so sorry for how hard this is for you.
- **Attend to the concern:** I'm glad we are protecting him from the cause of his discomfort.
- **Redirect to an achievable goal:** His face looks so relaxed, do you have the same observation?