

# **Let's Have a Conversation! Using AAC to Improve Communication for Individuals with Rett Syndrome**

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## **My Goals**

- Share information about Rett Syndrome you may not know
- 3 key areas address to improve communication
- Show tools and strategies can use today to start expanding communication
- Show you what individuals with RTT are communicating using eye gaze technology – so much more than what people have known or expected them to do
- Importance of engaging individuals in conversation – moving beyond choice making, expression of needs and wants and giving them a means to interact socially



## What is Rett Syndrome?

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- “Rett Syndrome (RTT) is a rare non-inherited genetic postnatal neurological disorder that almost exclusively occurs in girls, but can be rarely seen in boys”
  - Rettsyndrome.org, 2014
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## Neurological Foundation

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- MECP2 – gene mutation in Rett Syndrome; postnatal, not degenerative
  - MECP2 is dormant, inactive in utero – so born with brain of a typical infant
  - Have all the neurons that migrate to the correct locations of the brain; there is no effect on the brain at birth;
  - MECP2 gene affects dendrite growth and connections between the neurons; no evidence of cell loss
  - Synaptogenesis rapid between 2 months to 2 years
  - Synapses – connections between the neurons – plasticity of the brain; can change and strengthen connections between the neurons
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## Research at a rapid rate gives hope for a cure

- Gene identified in 1999
- Insight into the function of the gene
- Rett syndrome is **not degenerative** 2005
- Mouse model developed 2007
- Symptoms reversed in the mouse 2010
- Insight into the causes of the regression and its stabilization ?inflammation ?neurotrauma ongoing 2012
- Drugs that showed improvement in mice are in clinical trials on girls
- Neuroplasticity or formation of new synapses is improved with therapies

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## Enriched Environment

- Important research on neurotrauma has implications for Rett syndrome
- New synaptic connections can be built
- Neuroplasticity: The human brain is always remodeling allowing for learning
- Studies with mouse models of Rett syndrome support the value of a varied and stimulating environment

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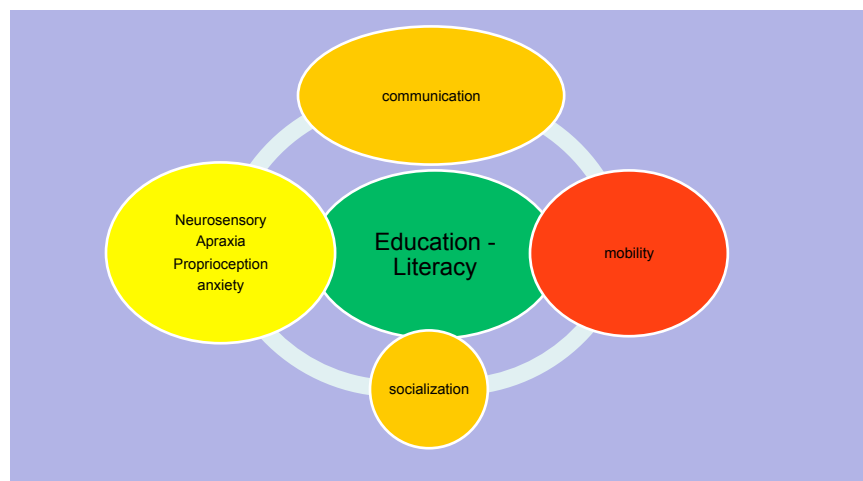
## How does “plasticity” apply to girls?

- Learning entails strengthening connections between neurons by creating more connections and enhancing ability to communicate and learn
- “Neurons that fire together, wire together” – Djukic, 2009
- Active engagement and participation is essential
- Emotional relevance – motivation and interest
- Stimulate thinking and imagination
- Girls are always achieving the expectations I set and beyond with instruction and strategies to support learning

The enriched environment for  
Rett syndrome  
includes elements that help her to  
access her education

- ☐ Communication and Literacy instruction
- ☐ Mobility
- ☐ Attention to apraxia, anxiety, neurosensory regulation
- ☐ Age appropriate socialization
- ☐ Medical issues

## Enriched environment



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## Natural history study of 1000 persons

- Showed different mutations have certain characteristics
- Showed that we cannot judge one girl with Rett Syndrome by experience of another – each is unique
- Patients who have had the benefit of therapies have fared better than girls born in earlier decades
- Life expectancy can extend well beyond middle age

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## Scored standardized testing is not an accurate estimate of cognition

- Testing is frequently done on entrance to school. On usual standardized tests children with Rett Syndrome will not score well as these tests are not normed on children with both loss of hand use and speech. Unfortunately in some instances the score is used to imply cognition, thereby missing the child's true learning potential.
- Individuals with Rett syndrome are interested and sensitive. Their physical tasks are great and when they are do not feel supported or valued it is difficult for them to summon the effort to try.

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## Unique Characteristics

- Stereotypical repetitive hand movements – involuntary
- The repetitive hand movements are pretty constant – you may need to hold one of her hands to break up the pattern and help with her focus
- Breath holding and hyperventilation
- Sensory processing difficulties – affects ability to attend, focus and participate, and give a motor response

## **Communication**

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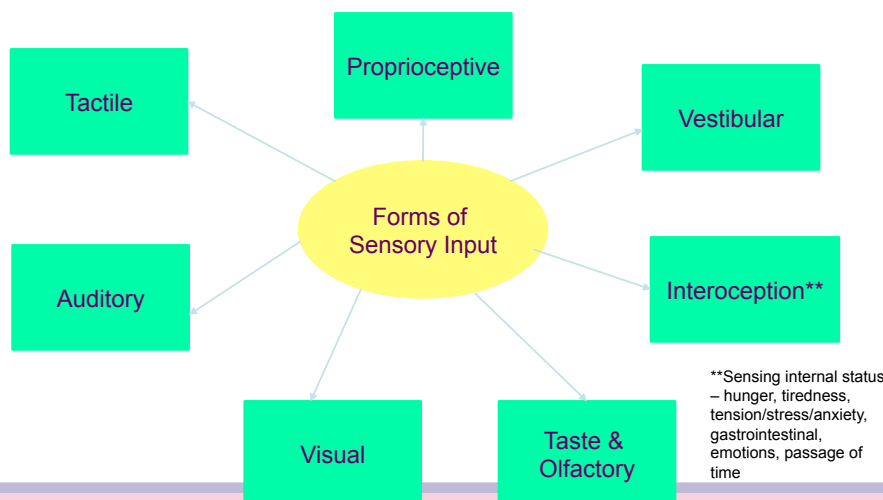
- Express oneself; open-ended
  - Not a test or only choice making
  - Includes building connections and strong emotional bonds with family members, including extended family, and friends
  - Want friendships, not just with adults, but same-age peers and siblings
  - This is possible through use of use of a variety of types of AAC options, including independent access to a communication device – their “voice” that they use with their natural gestures or unaided modes of communication
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## **Recipe for Enhancing Communication – Ingredient #1**

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- Monitoring responses to sensory input – making sure they are in a good zone and “ready” to communicate and participate
  - If they are not, their observable behaviors (i.e., fussing, walking around, or closing eyes/falling asleep) may be misinterpreted as inability to do or “refusal” to work
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## Forms of Sensory Input



## Proprioception

### Proprioception

This inability to filter or process information causes difficulty figuring out the body's position in space.

Proprioception translates to  
fear of being moved, fear of walking on uneven surfaces, fear of sudden movements



## Proprioception

- Training for all
- Promote seating balance
- Physical prompts for walking
- Balance for steps and uneven surfaces
- She leans to seek balance
- Unhurried transitions save time in the long run
- A misstep reinforces fear of movement, positive repetitions teach confidence

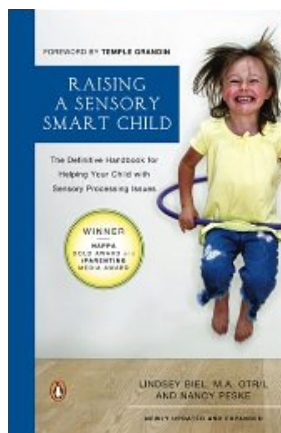
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## Processing of Sensory Input

- Each individual's response to various forms of sensory input is unique – avoid or seek/crave or neutral
- Parents know what makes their child upset or happy – clues to processing of sensory input – close links to behavior
- If possible, evaluation by an Occupational Therapist who specializes in assessing and treating Sensory Processing Disorder
- Develop and implement a sensory diet for home and school
- Intervention to improve how handle and respond to sensory input in various environments

## Recommended Website

- <http://sensorysmarts.com>
- Raising a Sensory Smart Child: The Definitive Handbook for Helping your Child with Sensory Processing Issues
- By Lindsey Biel, M.A., OTR/L and Nancy Peske (2009 edition)
- New Book by Lindsey Biel
- Sensory Processing Challenges: Effective Clinical Work with Kids & Teens



## Responses to Sensory Input

- Walks independently

Levels of Sensory Regulation	Description
Level 3	Distressed, agitated, refusal/ resistance to participate
Level 2	Difficulty remaining seated; walking around aimlessly,
Level 1	Calm, attentive, interested and ready for engagement

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## Responses to Sensory Input

- Requires assistance to walk and/or stand

Levels of Sensory Regulation	Description
Level 3	Asleep or drowsy, eyes shut or agitated/crying/whining; unable to participate
Level 2	Cranky/fussy; closing eyes; not maintaining head upright position; fleeting or no eye contact
Level 1	Calm, alert and focused, head up, making eye contact and/or attention to activity at hand.

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## Sensory Regulation

<b>OVER STIMULATED Sensory Overload</b>	<b>Level 3</b>	Needs strong sensory input from sensory diet to "calm down," overwhelmed May also shut down; fall asleep
<b>OVER RESPONSIVE</b>	<b>Level 2</b>	Needs sensory break to help return to Level 1; snack or drink; music or change of activity
<b>FUNCTIONAL STATE OF AROUSAL</b>	<b>Level 1</b>	Ready to communicate and learn
<b>UNDER RESPONSIVE</b>	<b>Level 2</b>	Needs break involving movement/change of position; snack or music
<b>UNDER STIMULATED</b>	<b>Level 3</b>	Shutdown; falling asleep; needs strong sensory input to "rev up" system

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## **Video - Request a break!**

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## **Daily Communication and Learning**

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- Identify sensory input that is \*\*calming or alerting
- The girl needs movement breaks
- Always need to determine when a girl is ready to communicate and participate
- Sensory system needs to be in a “regulated state” or in a functional state of arousal to support learning and communication
- Calm, alert, focused, making eye contact, engage in back and forth interactions

## **Recipe for Enhancing Communication – Ingredient #2**

- Take into account an individual's apraxia when communicating, especially when using technology
- One of the biggest challenges
- From Rettsyndrome.org's glossary  
"Apraxia is difficulty with the usual automatic planning done by the brain to execute voluntary movements."

## **Apraxia**

- Individuals with Rett syndrome have difficulty making their muscles work for them.
- Apraxia is the term for difficulty with motor planning or difficulty carrying out cognitive intent.
- Slow response time
- Slow transitions
- Retropulsion
- A sibling said, " her body is physically confused."

## Apraxia

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- Interferes with their response to a request to do something upon command, especially out of context
  - Affects motor movements, including the muscles controlling their eyes when using technology
  - Does not mean they don't understand what you are asking them to do or just being "stubborn"
  - The message from brain to a part of "their body gets scrambled or takes longer" response times
  - "Do this" or "look at the..." – being directive
  - Video – Apraxia – affects eyes too!
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## Apraxia

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- When a girl initiates a movement based on motivation or strong emotion, typically the effects of apraxia are reduced
  - This is often misinterpreted as the girls only want to do what they want to do; based on affective engagement
  - Video – 10 year old girl – trialing the new Tobii I-12; not seen or used my eye gaze trial page set for 8 months. **No evidence of apraxia – memory is a strength for the girls!**
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## **Apraxia**

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### **Strategies for working around apraxia:**

- Present/participate in motivating activities based on a child/teen/adult's interests or talk with them about a life experience
  - Say requests in such a way so they don't seem like a command/directive and wait quietly and watch....the gentle approach
  - Engage in natural and spontaneous interactions through play or conversations
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### **Recipe for Enhancing Communication – Ingredient #3 - Ease of Access**

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- Determine access first to a communication device
  - Must represent an easy motor plan; build on natural spontaneous movements and gestures
  - Avoid holding back communication development while working on hand use or waiting for other motor skills develop/improve or while waiting for eye gaze trials
  - Need 2,000 trials or repetitions in order for a child with complex needs to learn a new motor skill
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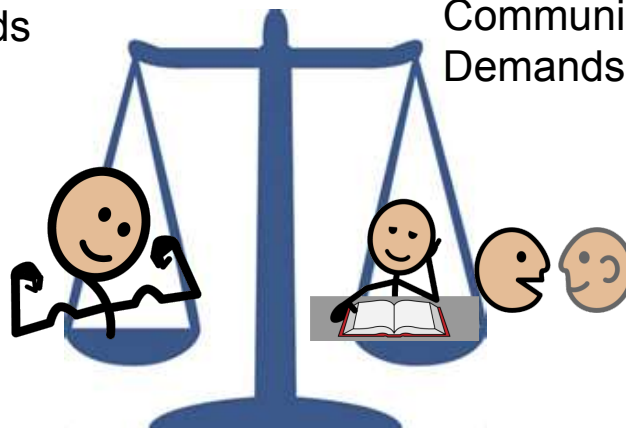
# **ACCESS is the key to unlocking potential in individuals with RTT**



## **Success - It is always a balance between...**

Motor  
Demands

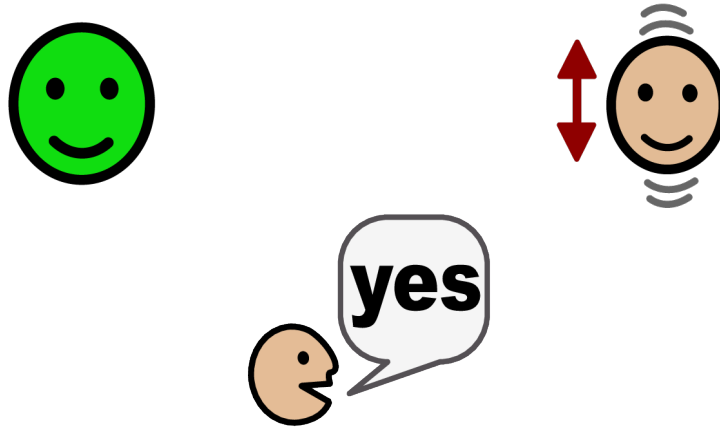
Learning and/or  
Communication  
Demands



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## Capitalize on “yes” response(s) using Partner-assisted scanning



## All individuals with RTT have natural “yes” responses/signals

- Need to observe to find out what it is; subtle
- In clinic, do you want to listen to music?
  - Wait and look for response – in most cases the girl will smile or smile and look at me; in some cases, eye brow raise, vocalize or slight head nod
- Build on natural “yes” responses; acknowledge these and shape them to increase frequency of use, consistency, and ease with which a girl uses it
- Absence of “yes” response can mean “no” or “I don’t know”
- Integrate it into partner-assisted scanning with communication-based app on the iPad

## Partner-assisted Scanning (PAS)

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- Present choices verbally and visually with a definite pause between and wait for the child to respond to indicate choice
  - Only need a “yes” response or a signal from child – lowers motor demands – focus on interaction and get more engagement
  - Allows for you to present more than one or two choices at one time; expand on what can access with hands directly
  - Incredibly useful technique for all individuals with RTT; especially at school for teaching academic concepts and active engagement in learning
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## Communication Flipbook

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- Portable
- Goes in a purse, supports communication anytime, anywhere
- Use with partner-assisted scan and one of best “yes” responses
- Use Flip ‘n Talk from Mayer-Johnson ([www.mayer-johnson.com](http://www.mayer-johnson.com))



## Communication – AAC2Go iPad app – click Start (If a young girl – between 2 and 4 years old does not have an AAC device)

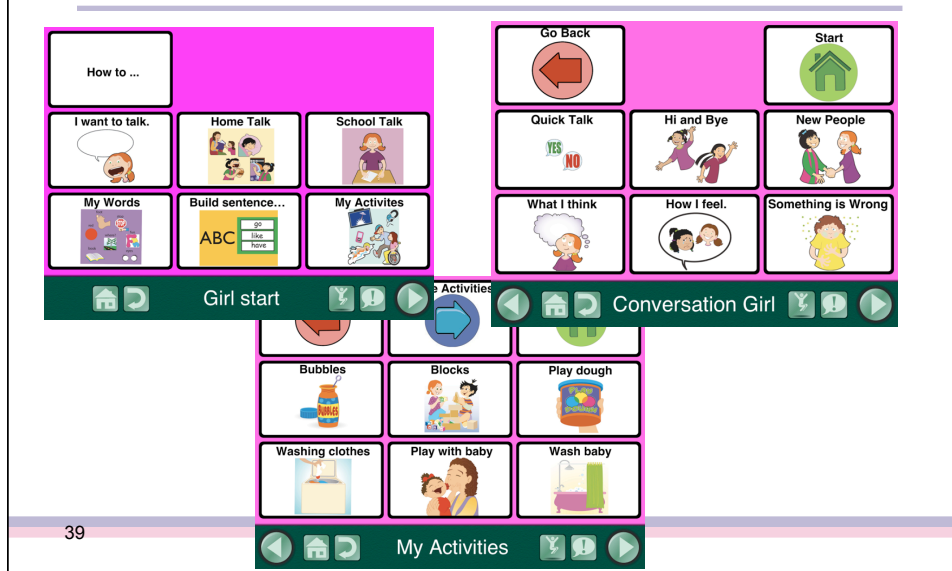


## Communication iPad – Go Talk NOW PLUS \$150.00

In-app purchase – AAC2go for girls – additional \$79.99 (also AAC2go Boys)



## AAC 2go for Go Talk NOW Plus App



39

## AAC 2go

- Easy to use and teach
- Quick success
- **Access to a “voice”**
- **Means of supported communication (does not replace need for independent means of access to “voice”)**
- Access to more than “wants and needs”
- For younger children based on the nature of the activities (i.e., bubbles, play dough, doll play, etc.)
- Vehicle for teaching literacy and language
- Pati is working on a teen/adult version (in beta)

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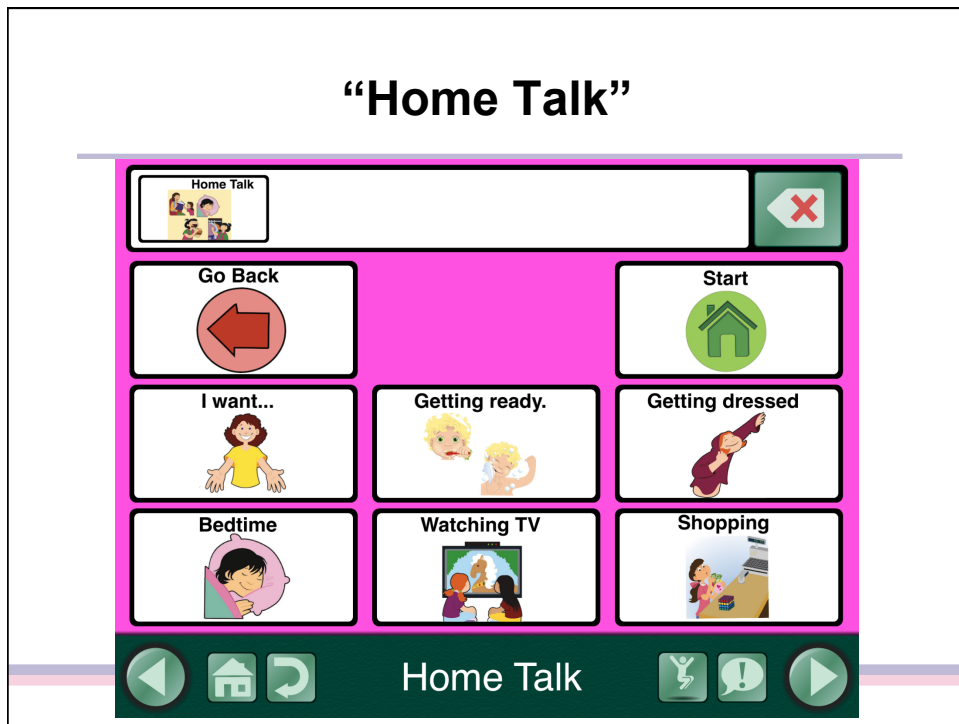
## “Something’s Wrong...”



## Success with AAC 2go

- Partner-assisted scanning
- One of girl’s best “yes” responses – (i.e., looking at communication partner when hearing desired choice named)
- Reacts to communication partner pressing the button on the iPad to speak the message in the same way as if she activated the button to play the message
- Access represents a challenge for a girl to access this many buttons on a page but she understands and can use these communicate.

## “Home Talk”



## “I want...”



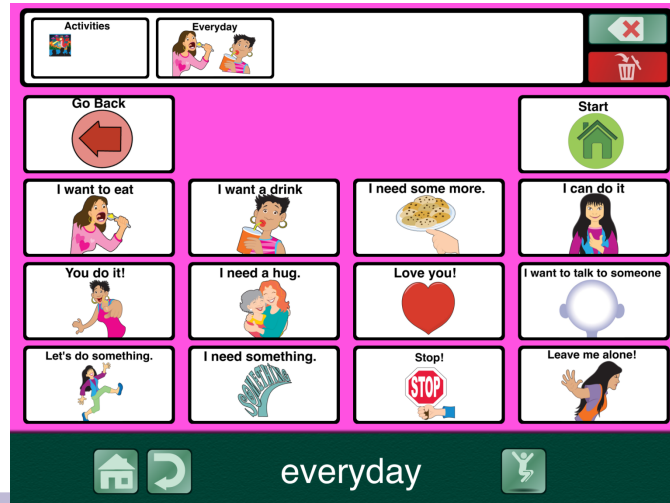
## 2 year old – first time using AAC2Go

- Quickly looked at communication partner when “I want to eat” and “I want a drink” to spontaneously indicate choices when these were named and pointed to by her communication partner

## Beta Version - Teen AAC2Go in-app purchase GoTalk NOW Plus



## Everyday

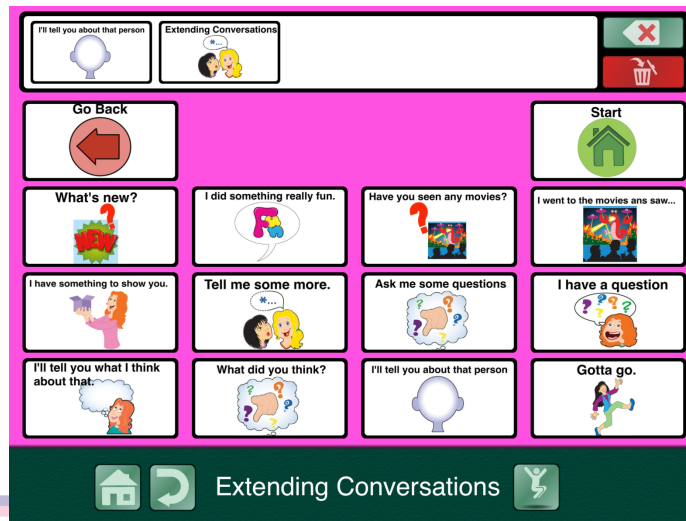


## Phone Talk





## Extending Conversations



## I'll tell you about that person

