

The Brain: Plasticity and Maturation In Sotos Syndrome

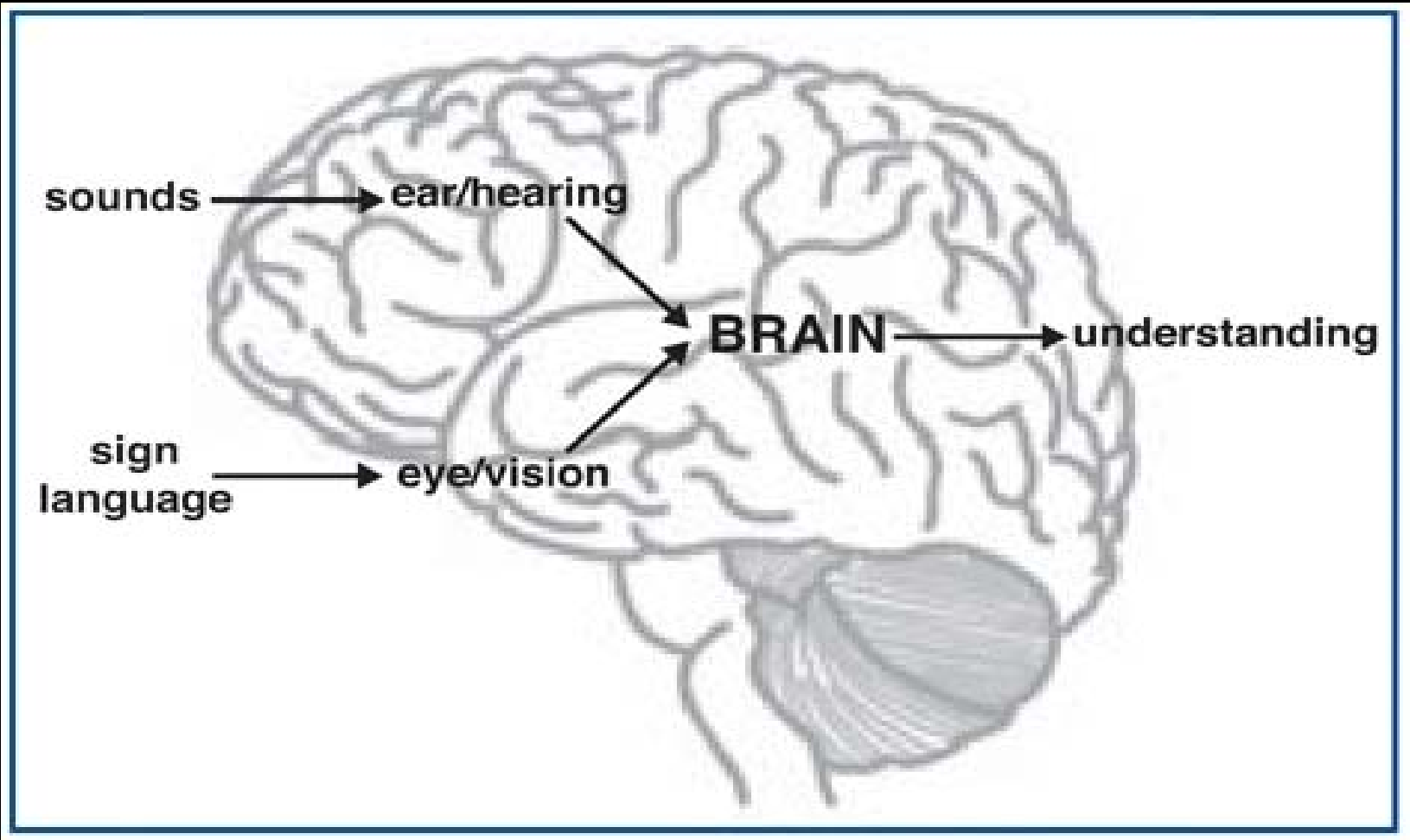
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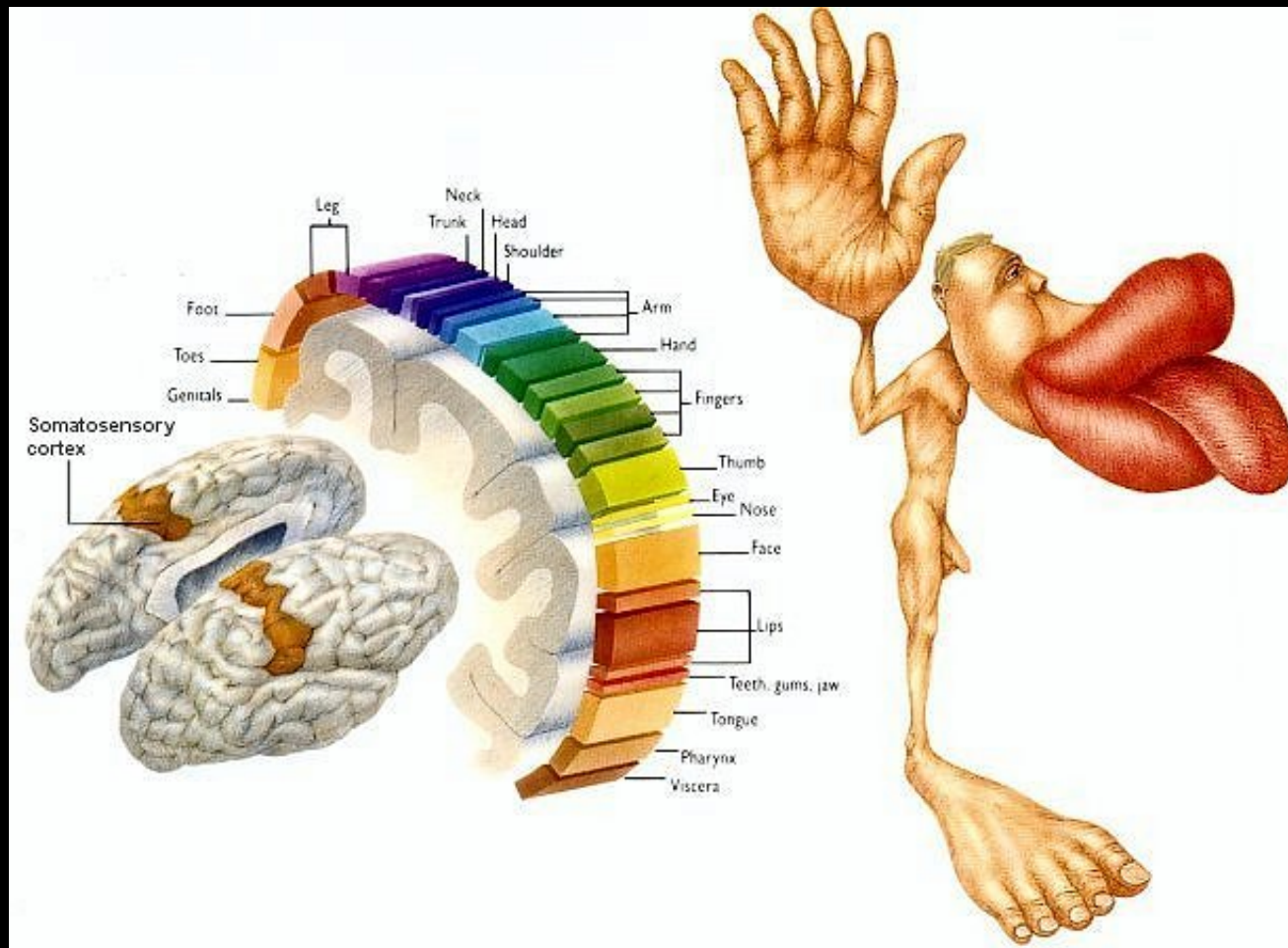
Brain Plasticity

Also known as neuroplasticity, is a unique characteristic of neuronal cells. No other organ has the capability of rewiring itself to take on other functions

Input increases cell synapses ----Leads to New Functions



Each part of the Brain has Unique Functions



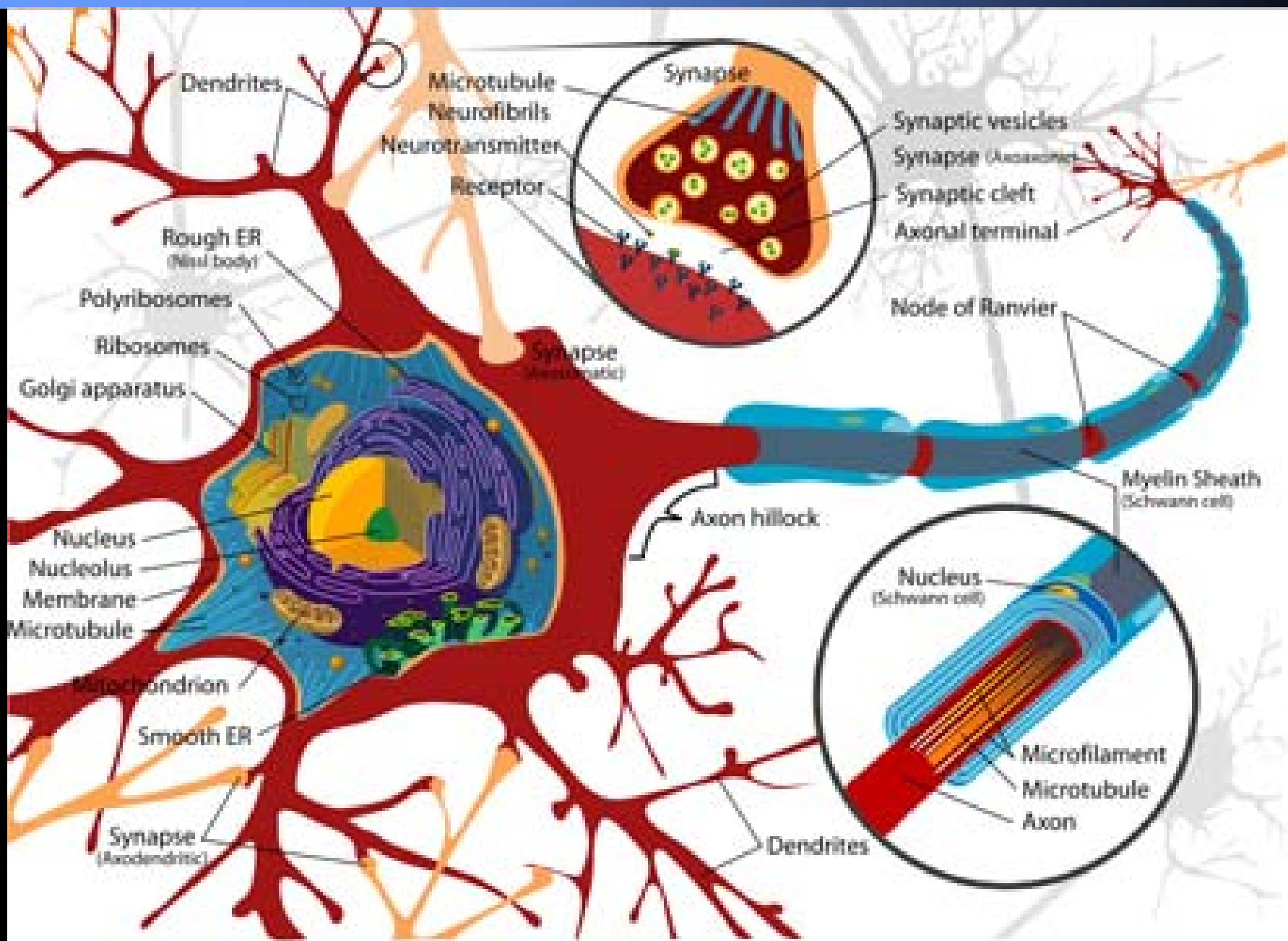
Maturation

Neural cells and myelin are added as the brain matures

- Directed motor and cognitive input directs the new cells to perform needed tasks (plasticity)

Neuronal Cells

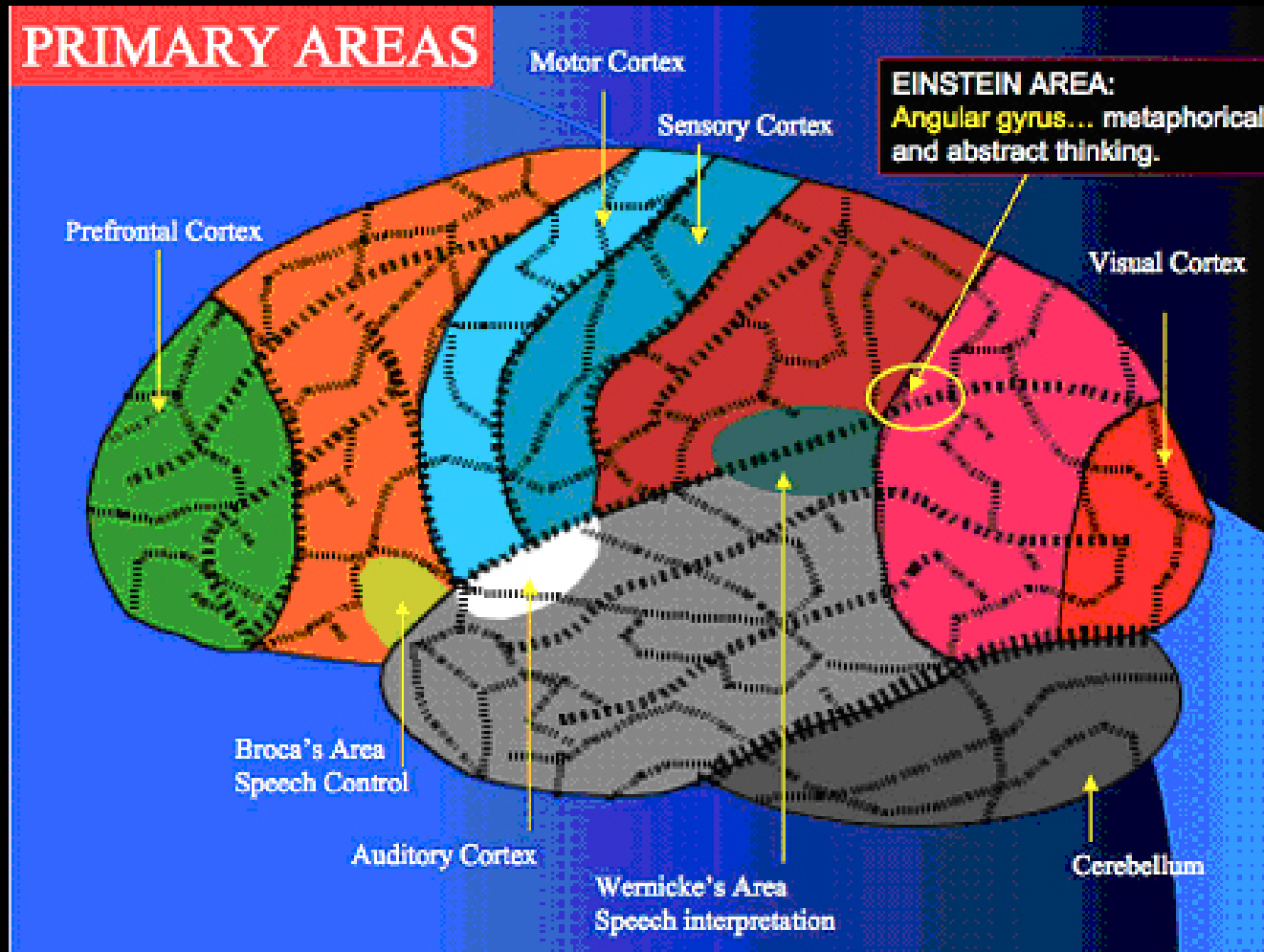
- *The brain constantly lays down new pathways for neural communication and rearranges existing ones throughout life, thereby aiding the processes of learning, memory, and adaptation through experience and repetition.*



Maturation of the Brain

- A continuous process
 - Myelination with definition of white-grey
 - Growth in Cortex (increased thickness)
 - Loss of specific neurons (pruning in adolescence and adults)
 - Specific areas of the brain mature at different rates

Functional Areas of the Brain

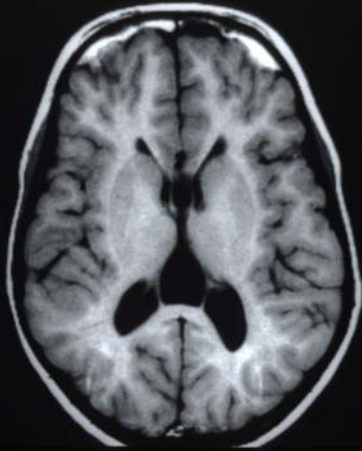


MRI Findings in Sotos Syndrome

- Neuronal Migration abnormalities
 - Delayed white-grey matter differentiation
- Ventricular Enlargement (not hydrocephalus)
- Abnormal Corpus Callosum
- Increased extracerebral fluid spaces

G. Bradley Schaefer et al

MRI Changes



- Ventricular abnormalities – prominence of the trigone (90%), prominence of the occipital horns (75%), ventriculomegaly (63%)
- Extracerebral fluid space enlargement - supratentorial (70%), posterior fossa (70%)
- Midline abnormalities – corpus callosum anomalies (nearly universal, most common – thinning of the middle 1/3)

MRI and Physical Correlations

Sotos Syndrome

- Hypotonia....

Decreased or low muscle tone

Reduced postural tone

‘Floppiness’

Poor Suck

Poor reflexes



Low tone

Delayed Walking
Down turned mouth
Delayed articulation



Immature Social Behaviors

- Plays with younger children
- Difficulties in larger groups
- Behaviors 1-2 years behind chronologic age.....longer "terrible two's"
 - Tantruming
 - Not picking up social clues

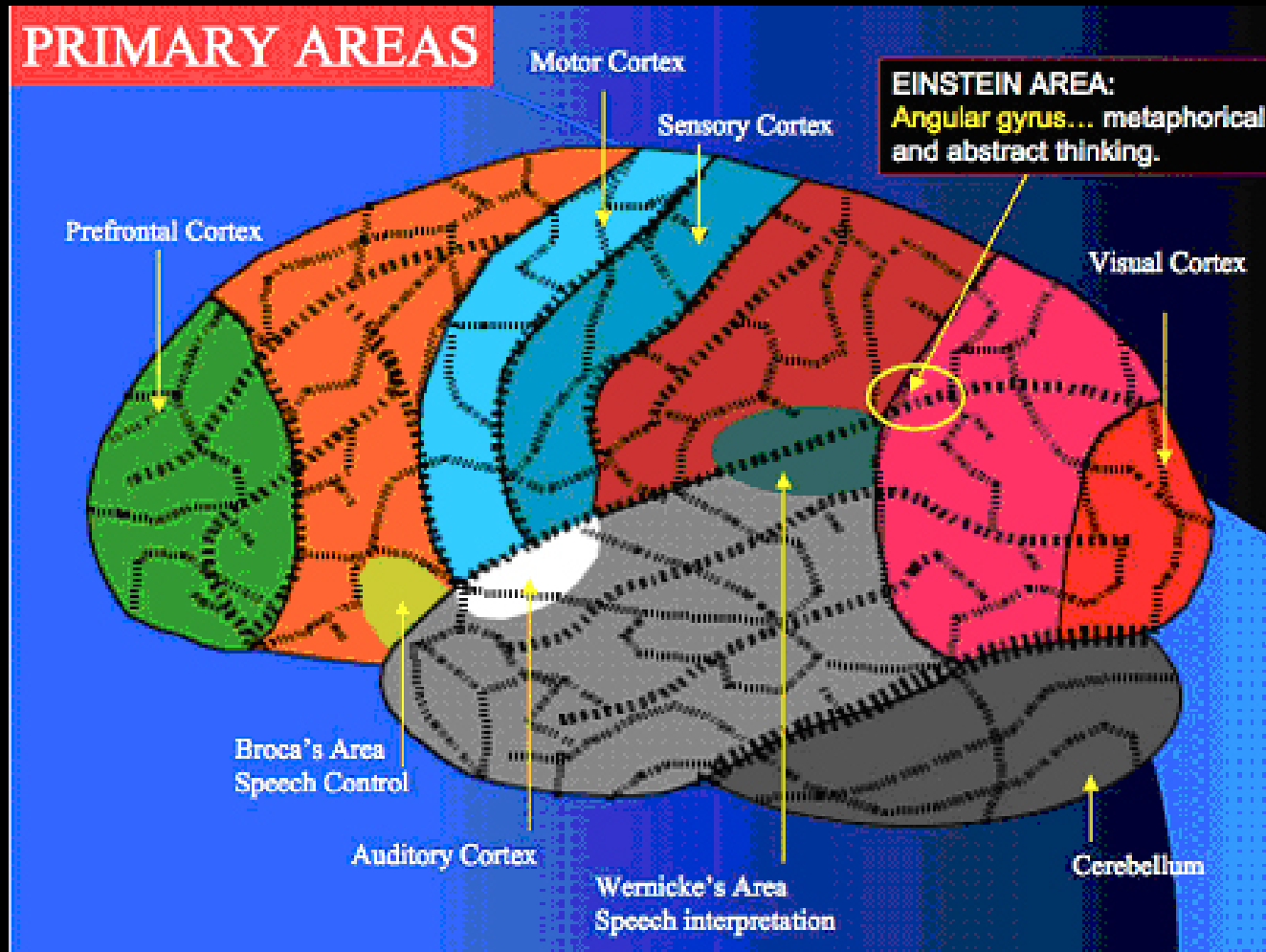
Speech Delays and Articulation

- Early speech delayed
 - May need sign language or pictures to develop an alternate pathway
 - Causes frustration
 - Requires repetitive input to enlarge Broca's area
- Articulation.....difficulties with hard sounds
 - Oral motor "training"

Brain Maturation continues in Sotos

- Gross and fine motor skills begin catch up around 10 years of age
- Improving "clumsiness"
- Social skills improve, "maturity"
- Behaviors become more attention difficulties and less frustration
- Speech catch up....
 - Improved articulation

Developing Brain



Plasticity and Maturation: Continuous

- Neurosensory integration

The integration of our visual, auditory, and balance systems. Occupational Therapy, Physical Therapy, Speech Therapy, and education

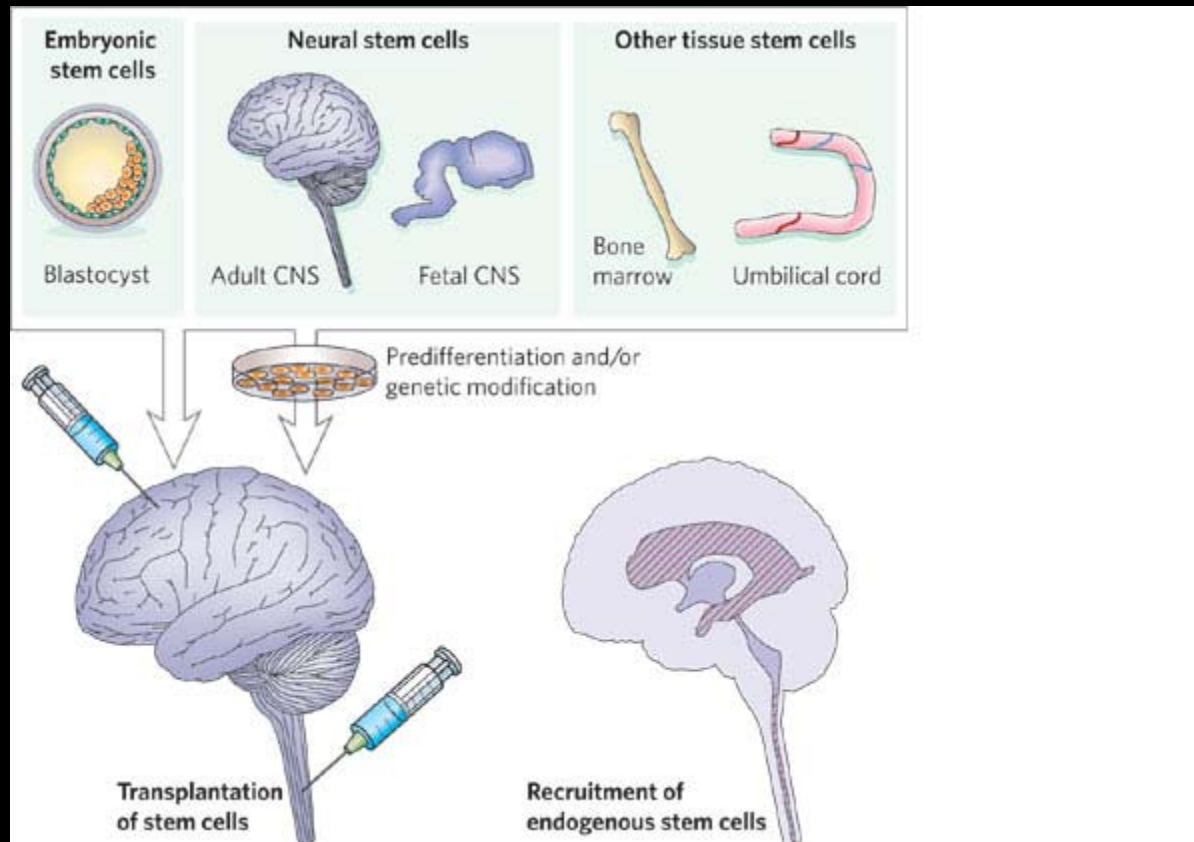
Learning Abilities
Sensory Processing
Memory, Organization,
Concentration

Plasticity and Maturation

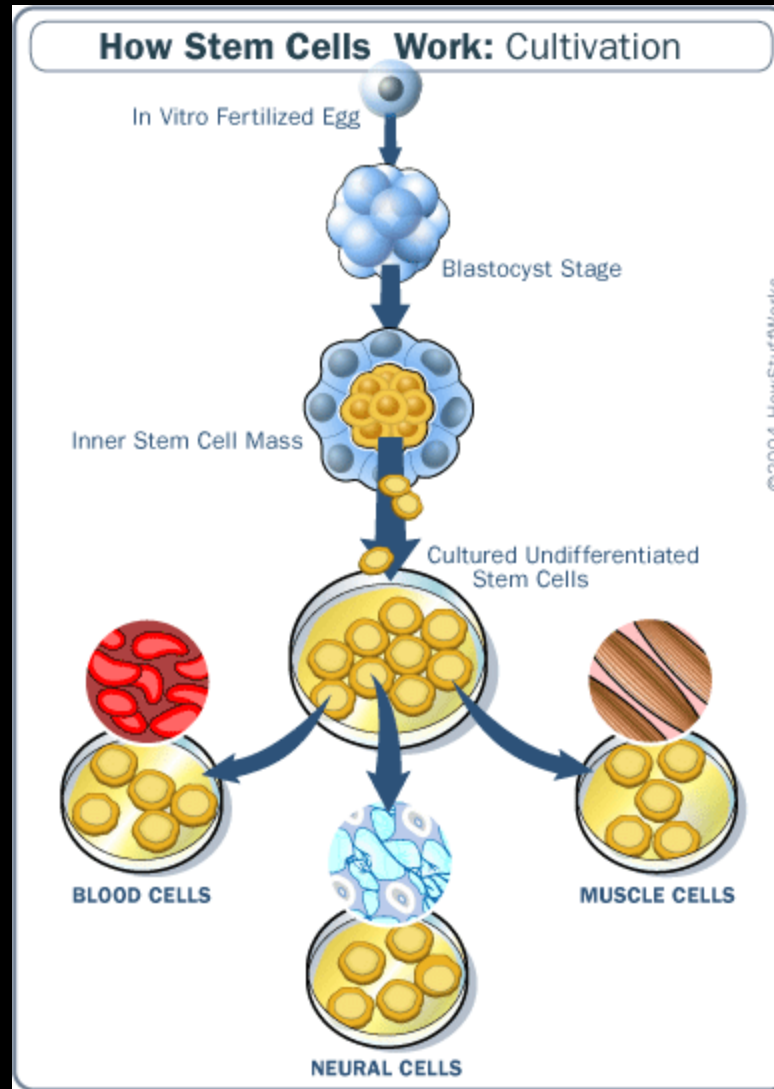
- Maturation increases the number of neurons.....both cognitive and motor
- All input (visual, auditory, and balance) creates new pathways, even developing alternate synapses to overcome areas of injury
- Damaged neurons are removed (pruning) and replaced



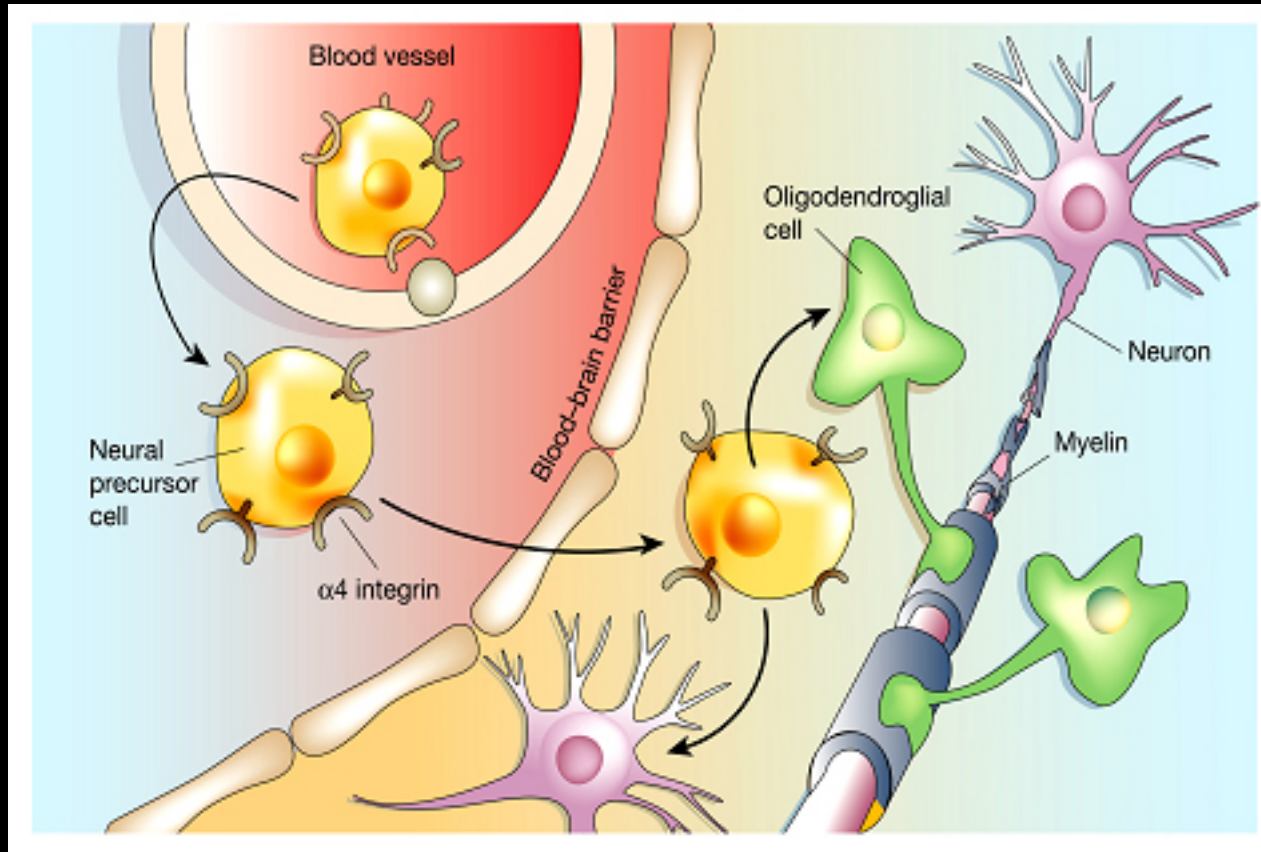
Research and Future Potential Stem Cells



Stem Cells



Potential for the Future



Questions?
Thanks.....Bruce

