



Speech and Language Therapy – a whistle-stop tour

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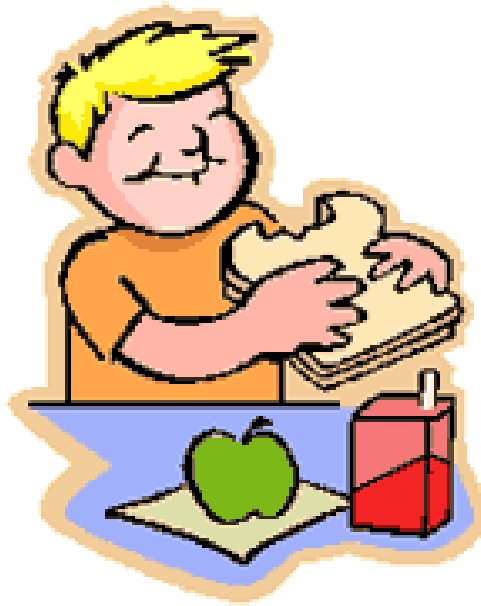


SLT as part of the clinical team

- All LSDs
- MPS III/Infantile Pompe predominance
- Feeding/Swallowing
- Communication

Eating and Drinking– the basics

safety

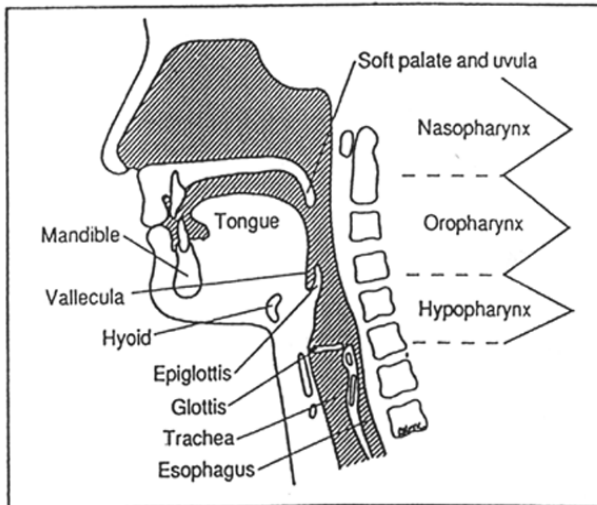
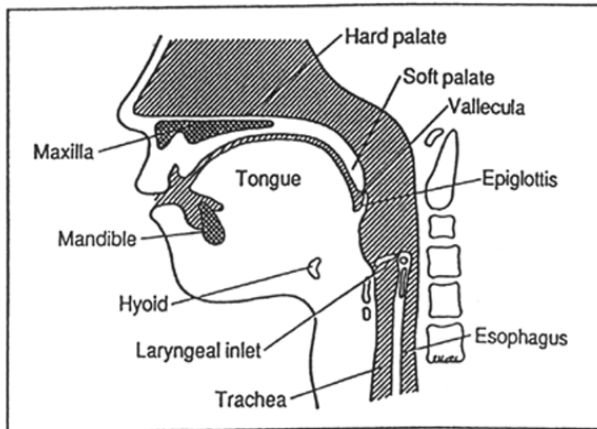


adequacy

social



The Mechanics



- Anatomy

- Infants

- Suck-swallow-breathe

- Respiration

Thinking about swallowing.....

- The normal swallow
- What happens when you swallow?
- Try swallowing with:
 - ◆ your head back vs chin down
 - ◆ Your mouth open
 - ◆ fast respiratory rate

Safety

- Airway protection
- Aspiration: *The entry of foreign material in to the airway, below the level of the vocal folds*
- Signs:
 - Colour change
 - Coughing/choking
 - Change in respiratory rate
 - Chest infections/aspiration pneumonia



Assessment – what do we do?

- History
- Watch
- Listen



Videofluoroscopy

*Image of videofluoroscopy
clinic*



Videofluoroscopy



videofluoroscopy

What about in Pompe?

- Jones et al (2009)
 - 100% some feeding difficulties
 - 77% airway invasion
 - 38% aspiration
 - No clear relationship between swallow dysfunction and overall clinical picture

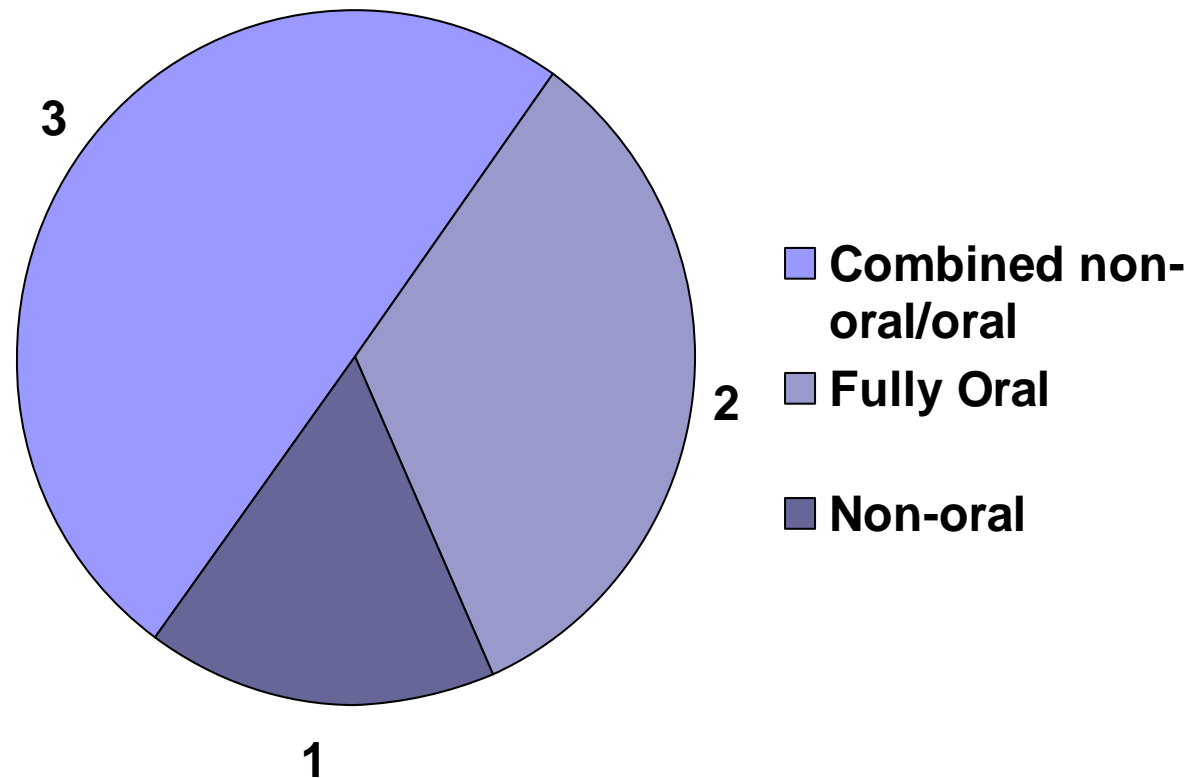
What can we do?

- Positioning
- Consistencies/Textures
- Nutrition management
- Positive experiences/desensitisation



GOSH caseload

- January 2011
- 6 patients



What is communication?

“The exchange of thoughts, messages, or information...Interpersonal rapport.”

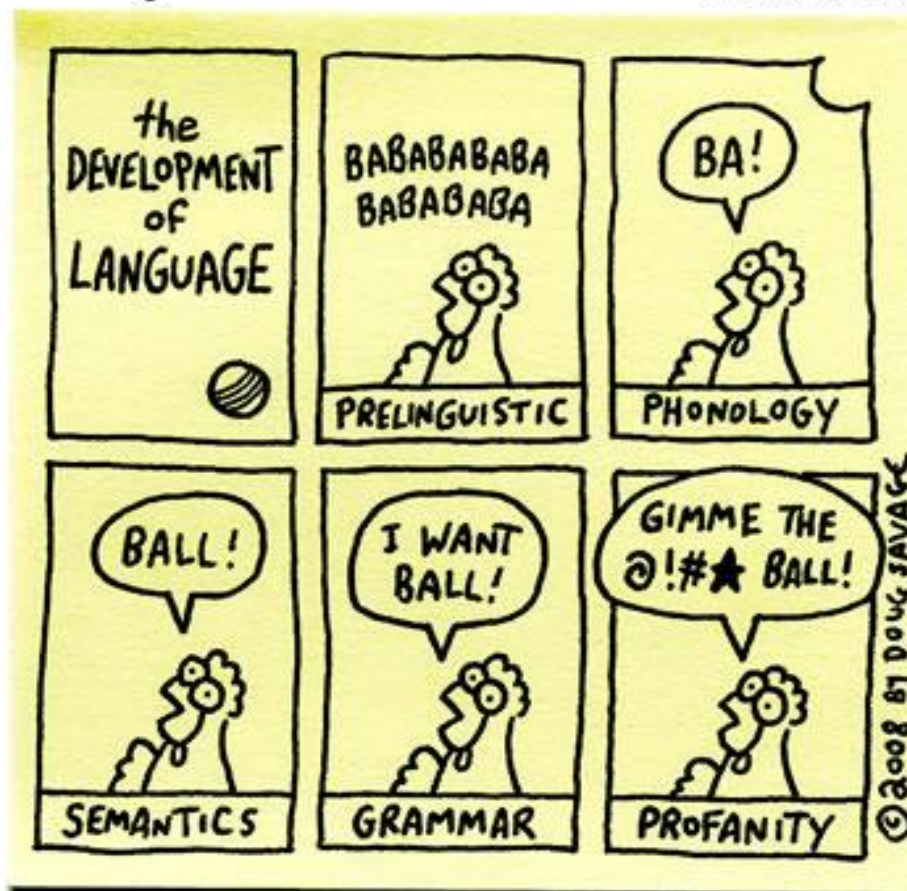
Thefreedictionary.com

- Language
- Speech
- Voice

How does children's language develop?

Savage Chickens

by Doug Savage

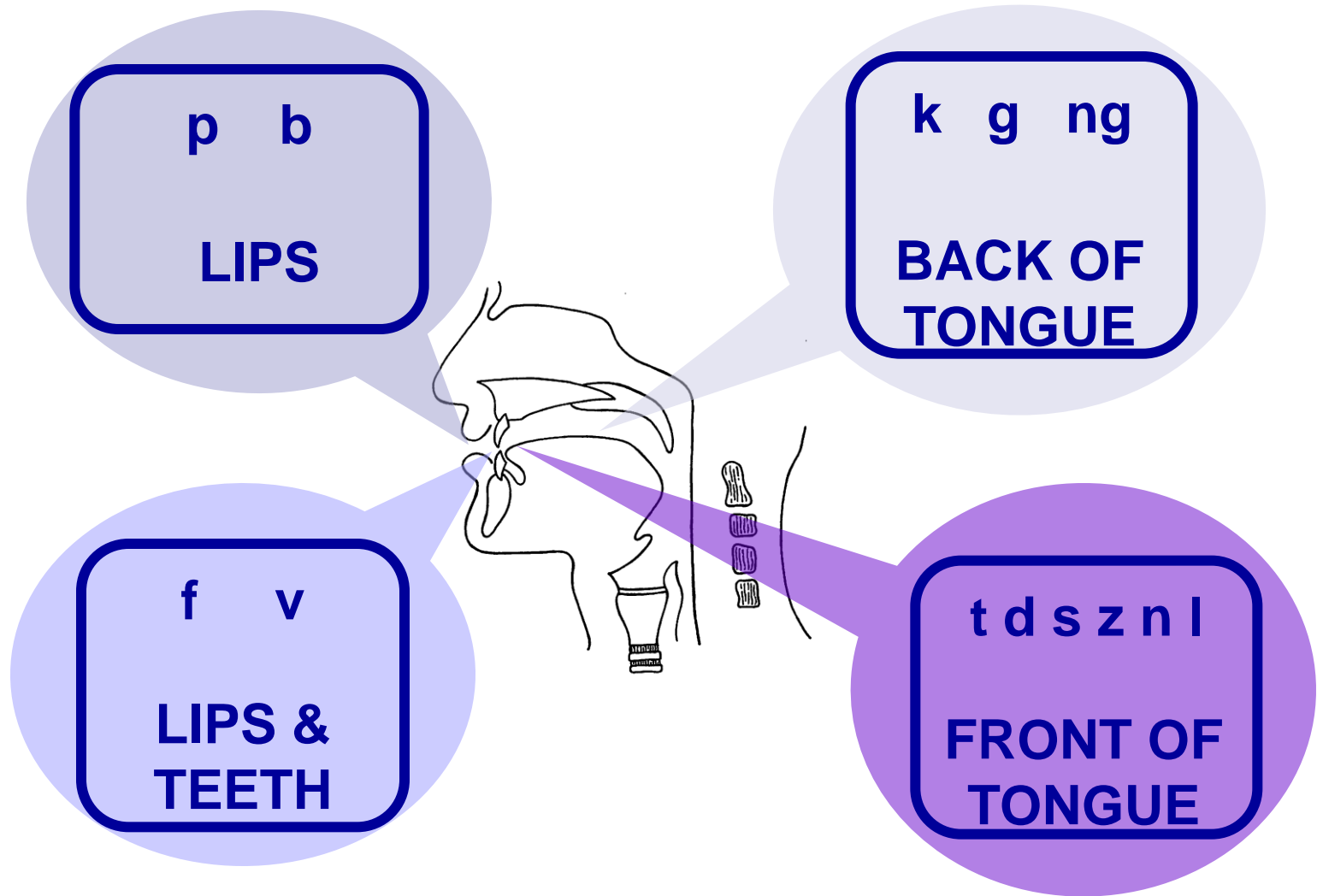




Speech

- Vocal fold vibration
- Modification of sound via
 - Oral/nasal airflow
 - Articulators

Where are sounds made?



What about Infantile Pompe?

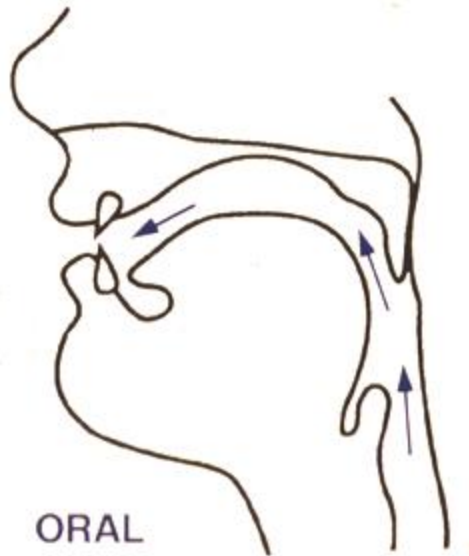
- Exciting time
- Muller et al (2009)
 - Language delay @ 18 months 42%
 - Reassessment @ 37months 83% resolved
 - Speech sound errors 82%
 - Hypernasality 82%

How the palate works for speech

Palate raised & touching back of throat.

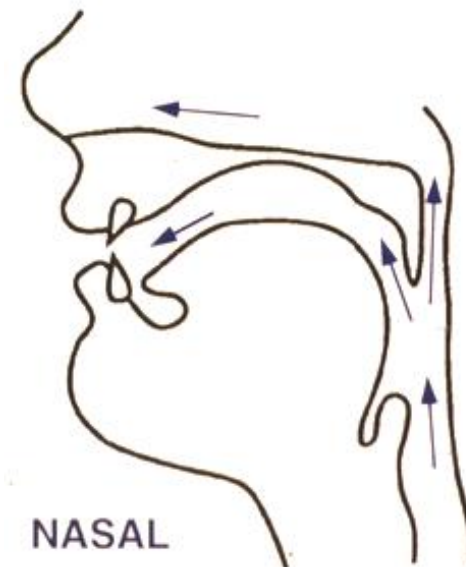
Air directed out of mouth.

Sounds: *p/b, t/d, k/g, f/v, s/z, sh, ch*



Palate raised but not touching back of throat. Air escapes through nose.

Sounds: *m, n, ng*





Lateral speech x-ray



Summary

- Emerging evidence
- Feeding and communication both affected
- Evidence of improvements in both fields.
- ?Natural history???
- ?Effects of ERT???