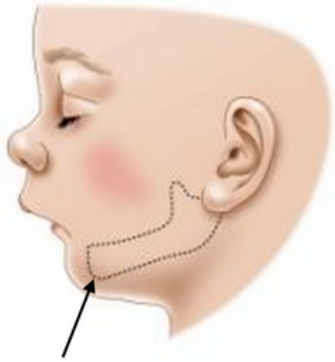
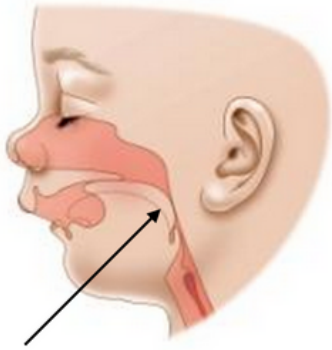
PIERRE ROBIN SEQUENCE AND FEEDING

What is it?

Pierre Robin Sequence (PRS) is characterized by a **small or underdeveloped jaw (micrognathia)** and a **tongue that is positioned further back in the mouth (glossoptosis)**. Airway obstruction is common due to the placement of the tongue and often, but not always, a cleft palate. Many children with PRS have co-occuring medical diagnoses like other genetic syndromes, heart conditions, and vision problems.



Micrognathia - a small jaw with a receding chin



Tongue that is large compared to the jaw, resulting in airway obstruction

Feeding Implications

70-90% of children with PRS have a U-shaped cleft palate (Dorise 2019) .
Babies with a cleft palate may find it difficult to latch onto a nipple and coordinate their sucking, making it challenging to obtain adequate nutrition.

Breathing Difficulties
This can cause fatigue during mealtimes, longer feeding times, and increased energy expenditure, resulting in a higher caloric requirement.

Feeding Difficulties
The structural abnormalities in PRS can increase the risk of aspiration (inhalation of food or liquid into the lungs) and frequent gagging or choking during feeding. Babies with PRS may also experience gastro-esophageal reflux disease (GERD).

Treatment and Strategies

Position changing when feeding and sleeping:
Strategies may include placing the child on their stomach or side to sleep and feeding in an upright position or side lying position to manage the airway obstruction.

Surgical intervention:
Common and effective surgeries may include: tongue-lip adhesion, tracheostomy, or mandibular distraction osteogenesis (MDO). MDO is the current technique to correct the airway obstruction (ASHA 2016).

Specialized nipple/bottle:
A child with may benefit from a specially designed bottle due to structural abnormalities to allow for easier access to nutrition and reducing aspiration.

Adjusting nutrition:
A child may benefit from strategies like pacing feeding, higher calorie formula, thickened liquid, or a feeding tube to meet nutritional needs.

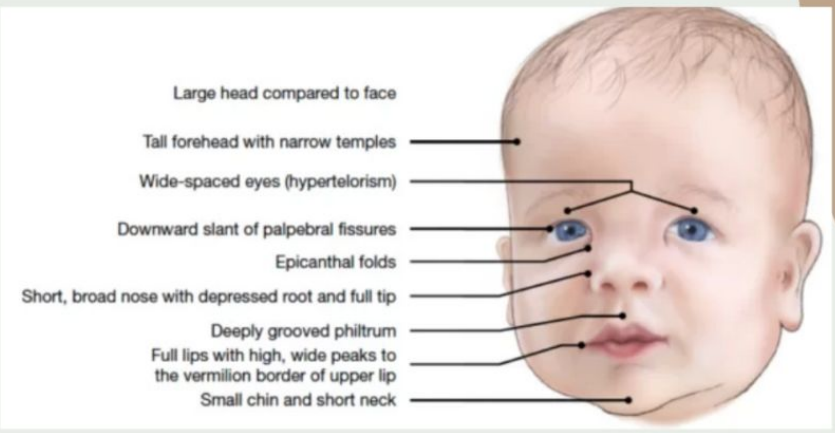
Resources for Families:

American Cleft Palate Craniofacial Association
acpacares.org/
National Organization for Rare Disorders
rarediseases.org/rarediseases/pierre-robin-sequence
Nationwide Children's Hospital
https://www.nationwidechildrens.org/conditions/pierre-robin-sequence#:~:text=In%20mild%20cases%2C%20Pierre%20Robin,through%20the%20nose%20for%20feeding

NOONAN SYNDROME AND FEEDING

What is it?

Noonan Syndrome is a congenital disorder that disrupts normal development in various parts of the body. It is characterized by distinctive facial features, short stature, and a range of congenital heart defects. Children with NS may experience developmental delays in areas such as walking, talking, and learning. This condition can also lead to other physical challenges, affecting overall growth and health.



Feeding Implications

"More than 50% of infants with NS develop feeding problems, and up to half of these infants will be tube-dependent for some time" (Tiemens 2022).

Frequent Vomiting

Gastro-esophageal reflux disease (GERD) and delayed gastric emptying is common along with a strong and persistent gag. This can lead to negative experiences with feeding.

Feeding Skill Dysfunction

Due to developmental delays, children with NS may have poor sucking and longer feeding times.

Oral Aversion/Sensory Needs

Children with NS are more likely to have texture sensitivities or lack a diverse diet.

Increased Caloric Need

Due to heart defects, children with NS may be expending more energy and need more calories to meet their nutritional needs.

Treatment and Strategies

There is a large improvement in feeding typically after age 2 (Draaisma 2020).

Modifying Dietary Volume and Variety

Modifying textures, blending foods, increasing calories in formula are strategies to meet nutritional needs. Chiuldren with NS may benefit from sensory or behavioral approaches to therapy to expand their diet.

Medical Management for Reflux

With the frequency of GERD and vomiting, medical management may be an option.

Therapy for Tube-Weening

Children with NS that are tube-fed can be seen by professionals to maintain oral feeding skills and to have assistance in tube-weening

Resources for Families:

Noonan Syndrome Association
https://www.noonansyndrome.org.uk

Noonan Syndrome Foundation
https://www.teamnoonan.org/

National Organization for Rare Disorders
https://rarediseases.org/rare-diseases/noonan-syndrome/

Turner Syndrome and Feeding

What is it?

Turner Syndrome is a genetic disorder that affects females due to a missing or incomplete X chromosome. It is characterized by short stature, hearing loss, high arched palate and displaced mandible (retrognathia), learning disabilities, and ovarian dysgenesis (poorly formed or missing ovaries).

Feeding Implications

Feeding Skill Dysfunction

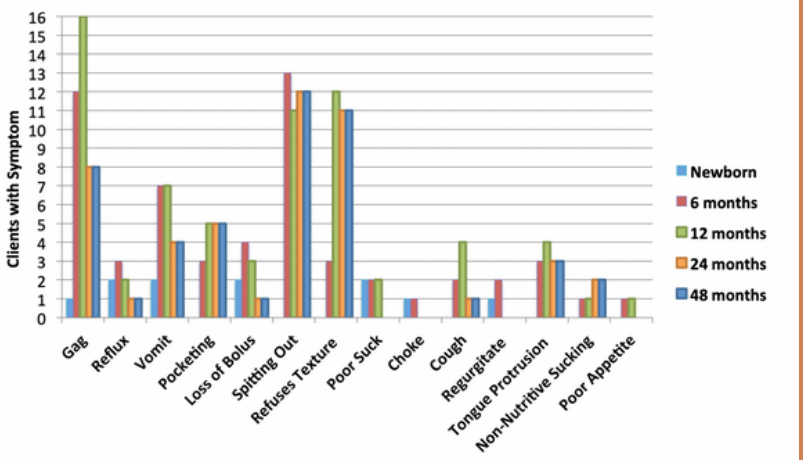
Due to a high palate, food may get stuck in the mouth or cause gagging. An abnormal jaw shape can lead to chewing difficulties.

Poor Oral Motor Control

Inefficient suck and swallow and dysfunctional tongue movement can cause difficulties obtaining nutrition and transitioning to solid foods.

Vomiting and Reflux

Girls with TS are at higher risk for Gastro-esophageal Reflux Disease (GERD) or delayed gastric emptying. This can lead to poor appetite.



Treatment and Strategies

Medical Management

Families can consult with a physician for medical management of reflux and vomiting. Babies born with TS may initially benefit from a feeding tube to obtain adequate nutrition.

Feeding Therapy

A child with TS may need therapy for transitioning to solids and practicing oral motor skills to increase jaw support, tongue coordination and chewing skills.

Specialized bottles and cups

Specialized bottles like the Haberman Feeder bottle can compensate for poor sucking skills. Small spoons and cups with thick rims may be easier for a child with TS to grip.

Limit Distractions During Mealtimes

Children with TS are more prone to hyperactivity and may become easily distracted during feeding.

Resources for Families:

Turner Syndrome Foundation

https://turnersyndromefoundation.org/welcome/

Turner Syndrome Support Society

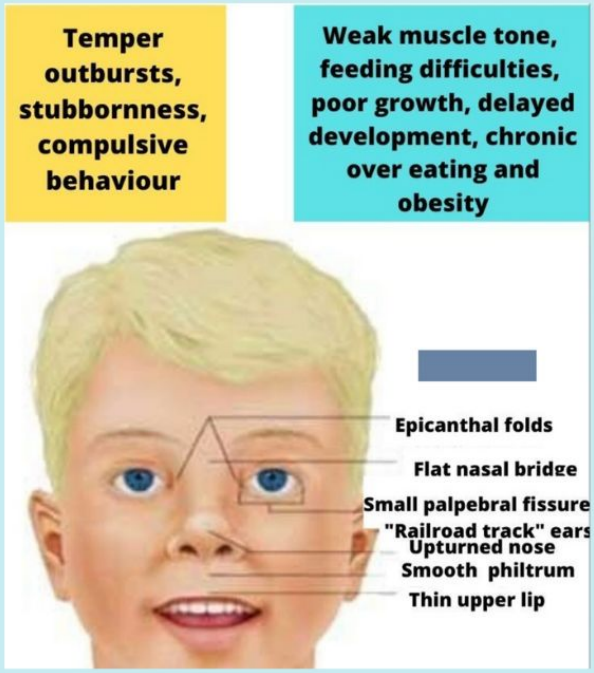
https://www.tss.org.uk

Child Growth Foundation

http://childgrowthfoundation.org/wp-content/uploads/2018/07/08_Turner_Syndrome_.pdf

In a study at the Carolina Institute of Developmental Disabilities, 41 participants with Turner Syndrome, ages 0-4, were categorized by feeding symptoms and age (Lam).

Prader-Willi Syndrome and Feeding



What is it?

Prader-Willi syndrome (PWS) is a genetic disorder caused by a deletion on chromosome 15, typically inherited from the father. It is characterized by intellectual disability, short stature, and constant hunger, which often leads to obesity. Individuals with PWS also experience behavioral issues and delayed puberty due to hormonal imbalances. The syndrome affects multiple aspects of development, requiring lifelong management and support.

Feeding Implications

Poor Suck-Swallow Response

Babies with PWS often do not have a strong suck-swallow response and may need to be tube-fed to meet nutritional needs temporarily.

Difficulty Controlling Food Intake and Risk for Obesity

Children with PWS expend less energy and need fewer calories. In addition, children with PWS may have increased hunger (hyperphagia) and poor impulse control.

Increased Risk for Aspiration

Children with PWS may have low muscle tone and motor planning issues that can lead to unsafe swallowing.

Treatment and Strategies

Manage Weight and Diet

Children with PWS benefit from diet support through a balanced diet, regular mealtimes, portion control, and restricting access to snacks.

Hormone Therapy

Treatment with the artificial Human Growth Hormone (HGH) is recommended for many children with PWS to manage their body weight, strength, and energy levels.

Oral-Motor Therapy

Therapy is recommended to improve sucking strength, oral motor skills for feeding, and to strengthen trunk and head support in order to swallow safely.

Resources for Families:

Prader-Willi Syndrome Association
https://www.pwsausa.org

International Prader-Willi Syndrome Organisation (IPWSO)
https://ipwso.org

Foundation for Prader-Willi Research
https://www.fpwr.org/what-is-prader-willi-syndrome

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