

Sorting Out Neurologic Swallowing & Feeding Problems in Infants & Young Children

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Prevalence of Feeding Problems

- 25% - 45% in children with normal development
- 33%-80% in children with developmental delays or chronic disease
- Up to 80% of children with cognitive deficits (mental retardation)

Etiologies for Feeding & Swallowing Problems in Pediatrics

Angelman Syndrome	Costello syndrome	Pierre Robin sequence
Severe atopy	Craniosynostosis	Eagle-Barrett syndrome
Autism spectrum disorders	Cri-du-chat	Robinow syndrome
Canavan syndrome	Dandy Walker Syndrome	Short gut
Cat eye syndrome	Diabetes	Spina Bifida
Chromosomal etiologies	Down syndrome	Stickler syndrome
Prematurity & complications	Eosinophilic GI disease	TEF
Orofacial malformations	Escobar syndrome	Solid organ transplantation
Airway malformations	Hirschsprung syndrome	Turner syndrome
Cockayne syndrome	Hemolytic uremic syndrome	VATER
Congenital diaphragmatic hernia	IUGR	Velocardiofacial syndrome
Congenital heart disease	Klinefelter syndrome	
Cornelia DeLange	Mitochondrial disease	Formula intolerance
	Noonan syndrome	Choking phobia
	Panhypopituitarism	Breastfeeding difficulty
	Cerebral palsy	"Sleeper eaters"
	Seizure disorders	Absent hunger drive

Neurologic Etiologies of Dysphagia

- Central nervous system (CNS)
 - Acute
 - Chronic
 - Static
 - Progressive
- Anterior horn cell (Infantile SMA)
- PNS (polyneuropathies)

Etiologies of Dysphagia (cont.)

- Neuromuscular junction (e.g., Myasthenia gravis)
- Muscles (e.g., congenital myopathies)
- Respiratory tract (e.g., BPD)
- Cardiovascular disorders (e.g., CHD)
- Gastrointestinal tract (GI)
- Psychological

Neuromuscular Disorders

Generalized

- Myasthenia Gravis
- Muscular Dystrophy
- Congenital Myopathies
- Cerebral Palsy
- Polymyositis or Dermatomyositis
- Infant Botulism
- Guillan-Barre

Oropharyngeal

- Arnold-Chiari Malformation
- Bulbar Atresia
- Ocular-Pharyngeal Dystrophy
- Brain Stem Tumor
- Drug-Tardive Dyskinesia
- Moebius Syndrome
- Cricopharyngeal Achalasia

Acquired Brain Injury

- Traumatic brain injury
 - Shaken baby syndrome
 - MVA, etc
- Seizure disorder
- Tumor
- Stroke

Other Disorders Associated with Feeding Difficulties

- Familial Dysautonomia
- Prader-Willi
- Hypothyroidism
- Trisomy 18 & 21
- Velocardiofacial Syndrome
- Rett Syndrome

Presentations of Feeding Disorders

- Inadequate growth due to inadequate intake
- Prolonged time for feedings (but with adequate calories for growth)
- Delayed progression of oral feeding skills (textures, variety, etc)
- Recurrent respiratory disease (question of aspiration from above or below)
- Complicating factors: behavior, sensory, relationship, social

CNS vs Neuromuscular Causes of Dysphagia in Childhood

- Passive tone
 - CNS – variable, hypertonia common
 - Neuromuscular – hypotonia
- Active tone (strength)
 - CNS – normal or mildly decreased
 - Neuromusc – Significantly decreased

CNS vs Neuromuscular Causes of Dysphagia in Childhood

- Primitive reflexes
 - CNS – usually strong & persistent
 - Neuromusc – may be absent or normal
- Plantar response
 - CNS – commonly up going (Babinski)
 - Neuromusc – plantar flexion

CNS vs Neuromuscular Causes of Dysphagia in Childhood

- Cognitive
 - CNS – deficits are common
 - Neuromuscular – usually normal

Cranial Nerve Deficits Associated with Dysphagia

- Supranuclear
 - V – mandible movements preserved, tonic bite reflex
 - VII – paralysis of lower half of face, usually unilateral
 - IX & X – normal strength in soft palate, vocal fold movement preserved
 - XII – Tongue movements present, dysfunctional; protrusion reflex may be exaggerated

Cranial Nerve Deficits Associated with Dysphagia

- Nuclear or Peripheral
 - V – Mandible movements minimal, absent tonic bite reflex
 - VII – Paralysis of upper & lower half of face – can be unilateral or bilateral
 - IX & X – Weakness of soft palate, vocal fold paralysis
 - XII – Unilateral or bilateral absence of tongue movements, fasciculations, atrophy

Chronic Aspiration: Pathophysiology

- May be more insidious than acute aspiration
 - Direct (primary, from above)
 - Indirect (secondary, from below)
- Swallow dysfunction & neuromuscular disease: most prone

Chronic Aspiration with Neuromuscular Disease

- Clinical indicators may be scarce
- Laryngeal penetration (level & frequency or consistency are important)
- Endangered airway
- Life threatening physiologic alterations

Pulmonary Disease with Neurologic Impairment

- Respiratory complications of dysphagia
 - Disordered timing/incoordination
 - Aspiration
 - Airway obstruction
- High risk infants (apnea & hypoxia)
- Older children: disorders of respiration
- Signs & symptoms of aspiration vary

Global Feeding Evaluation Goal

- To determine safest & most efficient consistencies for a child to eat orally (to whatever extent possible) while maintaining adequate nutrition & hydration

4 Key Questions to Ask Parents

- How long does it take to feed your child?
 - Longer than 30 minutes, tip-off for problem
- Are meal times stressful to child &/or parent?
 - Neurologic based skill & safety issues?
 - Behavior and/or sensory issues?
- Is your child gaining weight OK?
 - If no weight gain for 2-3 months, sign of problem
- Are there signs of respiratory problems?
 - e.g., congestion ↑ during feeding; gurgly voice

Goals of Clinical Evaluation

CAN'T

versus

WON'T

Development in Typical Child

- Liquid by nipple first 4-6 months
 - Breast milk
 - Formula
- Strained smooth food by spoon (6 months)
 - Sitting with minimal support
- Lumpy foods by 10-11 months
 - Difficult if delayed until 14-16 months
- Cup drinking before 12 months

Critical Period for Solids

Illingworth & Lister, 1964

- Spoon feeding by 6 months (developmental levels)
- Trunk support for sitting
- A-P tongue action reduced
- Hand-to-mouth skills
- NO mixed textures per bite

Age of Introduction to Solids

Age (months)	Type of Solid
4-6	Smooth puree (SP)
6-9	SP; Textured puree; Easily dissolvable solids
9-12	Soft, mashed, & diced solids
12-18	Toddler diet of chopped table food

Feeding/Swallow Evaluation

- History
- Physical examination
- Observation of typical feeding or mealtime
- Referral for additional examinations
 - Instrumental swallow study
 - Medical/surgical specialists
 - Nutrition
 - Psychology/Social Work
 - OT/PT

WHO Classification - ICF

- International Classification of Functioning, Disability, & Health (ICF): Potential framework for coding functional status & standardized language to describe & study health & health related domains. Further info at <http://www.who.int/classifications/icf/en/>
- First: Level of participation in various mealtime environments is considered

Holistic Approach To Evaluation

- Assists evaluators in decision making during treatment planning
- Focuses on overall goal of promoting a meaningful & functional mealtime experience for children & families

Steps in Clinical Evaluation

- Referral received ⇒ Initial Assessment
- Possible next step depends on airway status
 - If respiration normal, clinical feeding evaluation
 - If respiration abnormal, airway evaluation (hold feeds until airway is clear)

Clinical Evaluation: Airway Concerns?

- If none: Develop plan in context of global needs
 - Oral sensorimotor intervention
 - Nutrition guidelines
 - Behavioral therapy
 - Monitor status & alter plan as needed
- If yes: Instrumental examination or further medical workup

Feeding History Factors

- Positions/posture/seating (gross/fine motor)
- Duration of meal times (average & range)
- Intervals between meal times
- Types of food (preferred, non-preferred)
- Assistance/independence of feeding
- Tube feeding (e.g., type, timing)
- Food record: 2-3 days

Feeding History Factors

- Respiratory status
- Signs of stress & distress
- Test results & medications
- Sleep patterns (waking, snoring, mouth breathing)
- Cognition & communication
- Behavior during meals; apart from meals
- Therapeutic intervention (developmental/feeding)

Nervous System Exam

- Muscle tone
- Reflexes
- Cognition & language
- Visual tracking
- Gross & fine motor skills
- Sensory function

Infant Evaluation

- State & overall posture/positioning
- Respiratory status (rate, patterns, voice)
- Resting heart rate
- Exam of oral peripheral mechanism
- Non-nutritive sucking
- Nutritive suck/swallow/breathe sequencing

Evaluation of Transition Feeder & Older Child

- General observations
- Posture, alertness, direction following
- Oral sensorimotor function
- Bolus formation & oral phase of swallow
- Pharyngeal phase inferences
- Therapeutic trials

Cranial Nerve Evaluation for Feeding/Swallowing

- Lack of chewing: CN V
- Facial asymmetry & lack of lip movement: CN VII
- Delayed swallow & nasopharyngeal penetration/backflow/reflux: CN IX & X
- Tongue thrust or atrophy: CN XII

Gag Reflex

- Independent of swallow
- Sensory: CN IX
- Motor output: CN X, XII, & V
- Elicited by touching posterior pharyngeal mucosa (standard testing)
- Difficult to assess importance of changes in absence of other findings

Phasic Bite Reflex

- Rhythmic jaw opening & closing
- Response to pressure on gums
- Present by 28 weeks gestation
- Cranial Nerve V
- Disappears by 3-5 months
- May persist with neurologic deficit

Tonic Bite Reflex

- Jaw moves up into clenched position on presentation of spoon or other object
- Response to contact to biting surfaces of side gums (molar tables)
- Persistence with neurologic deficit – should disappear by 9-12 months
- Cranial Nerve V

Oral Sensory vs Motor Disorders

- Nipple confusion
- Inefficient suck breast & bottle
- Not differentiate tastes in bottle even with intact suck
- Differentiates tastes in bottle
- Manages liquids better than solid foods
- Oral-motor inefficiency or incoordination for all textures
- Sorts food in mixed texture
- Swallows food whole when given mixed textures

Oral Sensory vs Motor Disorders

- Holds food under tongue or in cheek and avoids swallowing
- Unable to hold & manipulate bolus on tongue, food falls out
- Vomiting only certain textures
- Vomiting not texture specific
- Gags when food approaches or touches lip
- Gags after food moves through oral cavity
- Hypersensitive gag with solids, normal liquid swallow
- Gags after swallow is triggered with liquid & solid

Oral Sensory vs Motor Disorders

- Tolerates own fingers in mouth, but not accept others
- Tolerates others' fingers in mouth
- Does not mouth toys
- Accepts teething toys, but not to bite or maintain in mouth
- Refuses tooth brushing
- Accepts tooth brushing

from Palmer & Heyman, 1993

Family & Child Interactions

- By direct observation & caregiver reports
 - Appetite, hunger, interest in eating
 - Regular meal times or snacks/grazing
 - Duration of meal times
 - Distractions needed
 - Rewards for eating

Immature vs Abnormal Patterns

- Children with immature oral skills are easier to manage than those with abnormal patterns
- Common for children to have both types

Immature vs Abnormal Patterns

- Patterns are likely to be distinguishable in
 - suck-swallow-breathe sequencing
 - jaw control or stability
 - tongue mobility
 - lip closure
 - dissociation of tongue, jaw, & cheek movements while drinking & chewing

Food Refusal

- Hyperextending head & neck
- Turning head away
- Spitting food out of the mouth
- Closing mouth tightly so food can't get in
- Pushing at sides of neck (pharyngeal residue?)

Next Steps?

- Nutrition Analysis
- Medical Workup (Genetics, GI, ENT, etc)
- Behavioral Psychology
- Occupational Therapy/Physical Therapy
- Instrumental Swallowing Study
 - Need to define oral, pharyngeal, & upper esophageal components for management
- Oral Sensorimotor Intervention

Criteria for Instrumental Evaluation

- Risk for aspiration by history or observation
- Prior aspiration pneumonia
- Suspicion of pharyngeal/laryngeal problem on basis of etiology
- Gurgly voice quality
- Need to define oral, pharyngeal, & upper esophageal components for management

Instrumental Swallow Evaluations

- Videofluoroscopic Swallow Study (VFSS)
 - AKA – Modified Barium Swallow Study (MBS)
 - Oral Pharyngeal Motility Study (OPMS)
 - Rehabilitation Swallow Study
 - Cookie Swallow + other names
- Fiberoptic Endoscopic Evaluation of Swallowing (FEES) + Sensory Test (FEESST)

Flexible Endoscopic Evaluation of Swallowing (FEES)

- No radiation
- Bedside exam possible
- Defines some aspects of pharyngeal physiology
- Can evaluate handling of secretions
- Sensory testing can be done

Videofluoroscopic Swallow Study (VFSS)

- Defines oral & pharyngeal phases
- Defines esophageal transit time & basic motility
- Delineates aspiration related factors
 - Before, during, or after swallows
 - Texture specificity
 - Estimate of risk

What VFSS is NOT

- To rule out aspiration or determine if child aspirates with oral feeding (important finding but not reason for exam)
- Simulation of a real meal
- Evaluation of oral skills for bolus formation
- Chewing evaluation
- Esophageal function (only upper esophagus)

Preparation of PO Feeders

- Hungry, but not starving
- Schedule close to feeding time if possible
- Normalize the situation as much as possible
 - Child's own utensils
 - Video/music as needed
- GT + PO: same guidelines as for total PO, unless child gets slow, continuous tube feeds

Preparation of Tube Feeder: NPO

- Child should demonstrate some level of oral intake, at least for therapeutic "taste trials"
 - NG tube – remove in some instances
 - Amount per bolus: 2 to 3 cc
 - Total of 10-15 cc preferred for validity & reliability
- Medication schedules maintained, or in some cases, adjustments needed

Child's "State"

- Typical feeding status appropriate
- Increased risks for aspiration
 - Lethargy
 - Agitation (fussing & crying)
- Cooperative child: interpretation possible in reliable & valid ways
- Always remember: Just a brief window in time, not a typical meal

Important Considerations in High Risk Pediatric Patients

- Radiologist must be present
- Well formulated questions
- Lateral view standard, A-P selective
 - Question of enlarged tonsils
 - Oral & pharyngeal asymmetry
- Audio important part of record
- Fluoroscopy time shortest possible while obtaining needed information

Procedural Decisions

- No fixed order for presentations in pediatrics
- Preferable to start with thinnest liquid
 - Controlled bolus size to start, e.g., spoon before going to bottle or cup drinking
- Work toward thicker as needed
 - Not want residue in pharynx that may complicate interpretation with thinner later
- Exceptions: parents tell us that child will not accept any thing else if he gets liquid first

Lateral View

- Encompassing
 - Lips anterior
 - Soft palate superior
 - Posterior pharyngeal wall posterior
 - Fifth to seventh cervical vertebrae inferior, varying with age of child
- Simultaneous view of oral, pharyngeal & upper esophagus before food is presented

Antero-Posterior View

- When asymmetry is known or suspected
- Unilateral vocal fold paralysis or paresis
- Tonsil related questions
- Other possibilities?
 - Keep in mind radiation exposure time
 - Importance of findings for management

VFSS Procedural Considerations

- Positioning/seating: typical & optimal
- Cooperative patient imperative for interpretation
- Caregivers included, findings reviewed
- Findings interpreted & used as part of total team approach: maximize safety
- Review in slow motion, frame-by-frame

Oral Phase Swallow Problems

- Lips (poor closing, drooling, leakage)
- Hesitation/pooling
- Tongue action deficits
- Gagging
- Poor posterior tongue thrust
- Passive leakage over tongue
- Delayed oral transit

Initiation of Pharyngeal Swallow

- Delayed swallow initiation/onset
 - Material in valleculae (not necessarily a delay
 - depends on timing)
 - Material in pyriform sinuses (has to be called a delay)
- Failure to initiate or trigger swallow

Pharyngeal Swallow Problems

- Nasopharyngeal backflow, reflux or regurgitation
- Penetration
 - To underside of epiglottis (superior)
 - To laryngeal vestibule/vocal folds
- Aspiration (below level of true vocal folds)
 - Response (or lack) to aspiration
 - Clearance of airway

Pharyngeal Swallow Problems

- Tongue base retraction reduced
- Pharyngeal contraction reduced
- Pharyngeal motility reduced
- Post-swallow residue, e.g.,
 - Valleculae
 - Pyriform sinuses
 - Posterior pharyngeal wall
- Clearance of residue?

Esophageal Swallow Findings

- Upper esophageal sphincter
 - Opening, e.g., reduced, incoordinated (often pharyngeal phase problem)
 - Prominence
- Bolus passage
 - Slow, interrupted
- Retrograde movement of contrast (better term than reflux in this instance)

Aspiration Before Swallow: Causes?

- Limited tongue action
- Limited mandibular movement
- Reduced tongue & soft palate approximation
- Delayed initiation/onset of pharyngeal swallow
 - Premature spillage
 - Material in valleculae & pyriform sinuses
- Pharyngeal dysmotility

Aspiration Before Swallow

- Most common with liquids
- Airway is open
- Neural control is voluntary

Aspiration During Swallow: Causes?

- Vocal fold paralysis/paresis
- Reduced laryngeal excursion
- Pharyngeal incoordination
- Nasopharyngeal backflow/reflux or regurgitation may be seen

Aspiration During Swallow

- Neural control
 - Initiation under voluntary control
 - Involuntary control for completion
- Airway
 - Closes upon initiation of pharyngeal swallow
 - Multiple levels of airway protection common

Aspiration After Swallow

- Reduced tongue base retraction
 - Residue in valleculae
 - Penetration into laryngeal vestibule
- Reduced pharyngeal contraction/motility
 - Residue in pyriform sinuses
- Reduced hyolaryngeal excursion
- Cricopharyngeal dysfunction
- Nasopharyngeal backflow may occur

Aspiration After Swallow

- Neural Control
 - Involuntary for esophageal phase
- Airway
 - Open
- Precipitating factors with open airway
 - Pharyngeal residue spills over
 - Nasopharyngeal refluxed material

Esophageal Dysphagia Diagnosis

- Dysphagia for solids > liquids, structural cause likely
- Dysphagia for solids & liquids similar, dysmotility likely cause

Interpretation of VFSS Findings

- SLP reviews with caregivers & therapists or others involved in care
 - Findings by phase of swallow
 - Timing of penetration/aspiration related to physiologic processes
- If review reveals a finding not anticipated or noted during exam, SLP contacts PA or radiologist to discuss or review together
- Important that reports are not discrepant

Problem Areas from VFSS

- Oral phase
- Initiating (triggering) pharyngeal swallow
- Pharyngeal phase
- Esophageal phase (upper)
 - Esophagram or UGI may be needed to define esophageal function

Report Outline

- Findings
 - By phase
 - By texture
- Impression or Interpretation
 - Correlate findings with physiology
 - Prognosis
- Recommendations

Management Recommendations

- Route for nutrition/hydration
- Feeding suggestions
- Therapy recommendations
- Additional suggestions
- Plans for follow-up or re-evaluation

Recommendations After VFSS

- Changes in route of nutrition/hydration
- Nutrition guidelines
- Position & posture changes
- Alterations of food textures, temperatures
- Utensil changes
- Changes in feeding schedule & pacing
- Oral sensorimotor program with food
- Nonnutritive oral sensorimotor program

Management: Prognosis & Priority

- Oral feeding prognosis tied closely to
 - Underlying etiology & diagnosis
 - Neurologic findings
 - Cardiopulmonary status
- Feeding priorities established on basis of
 - Severity
 - Combination of deficits

Principles for Repeat VFSS

- Same as for initial VFSS
- Information needed for
 - Definition of etiology or diagnosis
 - Guide for management decisions
- NOT some arbitrary time interval
- Child should be at baseline

Repeat VFSS: Primary Reasons

- Modification of current feeding routines anticipated
- Prior study lacks sufficient information for areas of concern
- Patient transfer & inadequate data

Evaluation Summary

- Comprehensive evaluation is vital
- Holistic approach is advocated for decision making (Functioning, Disability, Health)
- Social & physical mealtime environments considered along with oral skills & safety
- Goal is adequate health status; total oral feeding is not always a realistic goal

Instrumental Focused Summary

- Purpose & questions must be well defined
- Keep in mind: children with complex health & developmental issues may have many radiology studies throughout their lifetimes
- How will findings impact management decisions?
- A cooperative child is needed for reliable & valid interpretation of findings

Instrumental Focused Summary

- Remember: Study samples a brief window in time while the child is in an atypical eating situation
- Strive for development of noninvasive measures that can infer pharyngeal physiology so accurately that radiologic studies will not be needed. Children (& parents) will be happy.....

Neurologic Disorders with Poor Growth and Feeding/Swallowing Problems

Spinal Muscular Atrophy

- Second most common lethal autosomal recessive inherited disorder after CF
- Clinical features
 - Hypotonia
 - Muscle weakness (intercostal muscles)
 - Muscle atrophy
 - Fasciculations
- Degeneration of anterior horn cells, & variably, bulbar nuclei

Spinal Muscular Atrophy

- Symmetric muscle weakness, legs>arms
- No sensory deficits
- No intellectual impairment
- Gene defect: chromosome 5q (1990)
- Gene & its protein product, survivor motor neuron (SMN), were identified in 1995
- Diagnosis: gene mutation screening in 95%

Spinal Muscular Atrophy: Type I

- Werdnig-Hoffmann disease or severe infantile SMA – most severe
- Presents before 6 months
- Profound hypotonia, weakness, swallow dysfunction, tongue fasciculations
- Respiratory insufficiency
- Death by 2 years of age

Spinal Muscular Atrophy: Type II

- Intermediate or chronic infantile SMA
- Incidence 1/15,000 to 1/25,000
- Initially normal motor milestones that are lost during first 2 years of life
- Median age of death 12 yrs unless long-term mechanical ventilation
- Often not sit independently

Spinal Muscular Atrophy: Type II

- Weakness may be static for long periods
- Weakness progression during intercurrent illness or immobilization
- Bulbar musculature usually intact
- Nutritional evaluation & counseling needed due to poor weight gain from diffuse motor weakness leading to swallow dysfunction

Spinal Muscular Atrophy: Type III

- Kugelberg-Welander or mild SMA
- Onset between 2 & 17 years
- Usually able to stand & walk unaided

Guillain-Barre Syndrome

- Acute inflammatory ascending demyelinating disease of PNS
- Usually history of nonspecific illness
- Can be associated with cytomegalovirus & Epstein-Barr virus infection
- Ascending paralysis & weakness progresses over hours to several days
- Hypoventilation with diaphragm & intercostal muscles involved

Guillain-Barre Syndrome

- Progressive weakness involving cranial nerves: laryngeal & VF dysfunction with resulting upper airway obstruction
- Respiratory support required – serial monitoring of vital capacity & respiratory muscle function is critical
- Treatment: IV immunoglobulin, corticosteroids, & aggressive respiratory support
- Recovery usually complete from days to months

Myopathy

- Muscle weakness & wasting, often proximal
- Rare: Present with nutrition concerns
- Duchenne's dystrophy is exception
 - Most common lethal x-linked disease
 - Onset in early childhood
 - Consider with poor weight gain, motor delays, and muscle weakness

Duchenne Muscular Dystrophy

- Most common form of MD in children
- 1:3000 male births, X-linked recessive progressive neuromuscular disease
- Cause: mutations in dystrophin gene resulting in deficiency of normal dystrophin protein
- Gene localized to short arm of X chromosome (Xp21) in 1986.

Duchenne Muscular Dystrophy

- Boys present between 2-6 years old: frequent falling, waddling gait, toe walking
- Classic features: calf muscle pseudo-hypertrophy & Gower sign (climbing up legs using hands when rising from a seated position on the floor)
- Diagnosis made by muscle biopsy revealing missing or significantly deficient dystrophin.

Duchenne Muscular Dystrophy

- Becker MD – milder form
- Also affects dystrophin gene & protein
- Present at later age & walk up to early 20s
- Muscle biopsy: ↓ dystrophin concentration
- Genetic testing: identifies 60-65% of MD

Duchenne Muscular Dystrophy

- Motor weakness progression
 - Loss of ambulation by 11 yrs
 - Respiratory muscle function parallels gross motor weakness
 - scoliosis
- Cardiomyopathy contributes to early death
- Management: supportive, respiratory function

Duchenne Muscular Dystrophy

- Therapeutic intervention re swallowing
 - As weakness progresses, risk for aspiration increases
 - Fatigue becomes factor with weakness
 - Dietitian important to give guidance re maximizing nutrition, minimizing aspiration risks

Myasthenia Gravis

- Rare in pediatrics; primary disorder of neuromuscular transmission
- Postsynaptic receptors for acetylcholine functionally reduced in number by auto-immune mechanisms or abnormal protein formation
- Neonatal MG transplacental transmission of maternal acetylcholine receptor antibodies to neonate

Myasthenia Gravis

- Clinical presentation
 - Hypotonia, weak cry
 - Difficulty feeding
 - Facial weakness
 - Palpebral ptosis
- Respiratory compromise by aspiration with oral feeding

Myasthenia Gravis

- Resolves in 2-6 wks (neonatal type)
- Congenital MG: autosomal recessive, variable age of onset
- Diagnosis based on transient clinical improvement after administration of anticholinesterase medication

Microcephaly

- Associated with chromosomal disorders
- Profound nutritional deprivation without other conditions
- Associated with early PEM in first 2 years
- Changes in brain, especially 1st year
- Long term: reduced head circumference & significant cognitive impairment

Cerebral Palsy

- 1 child in 1,000 in U.S.
- Growth impairment by nutritional & other factors
 - Nutritional factors affect weight gain
 - Nonnutritional factors affect linear growth
- Severe impairment: multiple factors influence nutritional intake & risk for aspiration
- Group of CP: VLBW & complications of prematurity, e.g., CLD, and CPVL

CP: High Risk for Dysphagia

- Swallowing deficits worse with more severe CP
- Dysphagia & quadriplegia: independently associated with poor growth
- GT feeds: improve weight gain & correct undernutrition
- Development of nutrition plan depends on accurate assessment of feeding & swallowing

CP: Swallowing Problems

- Poor lingual function
- Tongue thrust
- Exaggerated bite reflex
- Tactile hypersensitivity
- Delayed swallow initiation
- Drooling
- Reduced pharyngeal motility

CP: Oral Feeding

- Solid boluses managed better than liquid
- Small liquid boluses easier than large liquid boluses
- Recommendations:
 - Children with CP need more time to complete feeding tasks (caution re prolonged meal times)
 - Small volume drinks better than large

Casas et al. 1994

Tumors of CNS

- May present with growth impairment without neurologic impairment
- Diencephalic Syndrome - symptom complex
- Posterior fossa tumors - recurrent vomiting
- Intracranial tumors: important to consider
 - Poor weight gain & growth +
 - Any CNS signs

Genetic Disorders, Growth Issues, & Dysphagia

Prader-Willi Syndrome

- Hypotonia followed by obesity in childhood
- 70%: deletion long arm of chromosome 15 at q11/q13
- Marked hypotonia in infancy, abnormal cry, & feeding difficulties
- Dx imp for prognosis & management

Prader-Willi Syndrome

- Poor fetal movements in utero
- MR & hypotonia
- Dysphagia common in first year, especially neonatal period
- Physical features
 - Pre- or post-natal growth retardation
 - Narrow bifrontal diameter
 - Almond-shaped eyes
 - Hypogonadism

Prader-Willi Syndrome Treatment

- In infancy, GT may not be needed, since prognosis is + for PO within first year
- Growth hormone (experimental treatment)
- Overall prognosis relates to combination of deficits & presence of other anomalies
- Current genetic research promising to improve understanding of etiology & course

Velo-Cardio-Facial Syndrome

- Microdeletion at chromosome 22q11.2
 - Incidence 1/5000
 - Autosomal dominant in most cases
- VPI & NP reflux
- Cardiac anomalies
- UES dysfunction common
- Hypotonia (pharyngeal in 90%)

Down Syndrome (Trisomy 21)

- Infancy
 - Hypotonia contributes to weak suck
 - Cardiovascular anomalies may contribute
- Degree of feeding difficulty likely relates closely to number & type of added problems

Down Syndrome

- Weight gain problems may be associated with cardiac, respiratory, & GI disorders
- Feeding difficulties may relate to upper airway obstruction, oral sensorimotor dysfunction neurologically based, food refusal, & chronic infections
- Growth charts for DS along with “typical”

Down Syndrome

- Problems relate to
 - Cardiac status - tire easily
 - Respiratory status - top priority
 - Hypotonia - weak suck
 - Asphyxia - incoordinated suck/swallow
 - Cleft palate in some

Trisomies 13 & 18

- Trisomy 13: 82% die within 1st month. Survivors: seizures, undernutrition
- Trisomy 18: >130 different abnormalities involving all organ systems
 - 50% die within the 1st week
 - Poor suck - supplemental feeds

Turner Syndrome (XO)

- Girls born with 1 of 2 X chromosomes partially or completely missing
- Strong association with short stature & poor weight gain
- Diagnosis often made at birth
- Suspicious physical features & poor weight gain associated with feeding difficulties: Consider Turner & get chromosomes

Turner Syndrome

- Newborns
 - Swelling on backs of hands & tops of feet
 - Swelling or loose folds of skin back of neck
 - Webbed neck, low hairline back of neck
 - Broad chest, wide-spaced nipples
- Older children
 - No menstrual periods, lack of puberty changes
 - Short & obese

Noonan Syndrome

- Inherited or unpredictable development
 - Gene localized to chromosome 12
- Normal chromosomal structure
- Characteristics typical of Turner syndrome
- Boys & girls – usually short
 - Intelligence may be impaired

Noonan Syndrome Intervention

- Growth hormone
- Testosterone treatment may help boys
- Feeding issues variable
 - Failure to thrive in some
 - No obvious cause for problems

Russell-Silver Syndrome

- Consider diagnosis:
 - Short stature, often prenatal onset
 - Triangular face
 - Asymmetry of limbs
- Close monitoring for hypoglycemia is needed

Fragile X Syndrome

- Genetic abnormality in DNA on the X chromosome (delayed development)
- Affected boys inherit condition from mother
- Intelligence
 - Many are normal
 - Most common genetic cause of MR besides Down syndrome (boys worse)

Fragile X Syndrome

- Poor weight gain may be associated with hypotonia & GER
- Consider: male infant or young boy with history of early undernutrition, vomiting, food refusal, & hypotonia
- Important clue: Family History of MR, especially when sex linked

Fragile X Syndrome Intervention

- Early intervention for speech/language, OT
- Stimulants, antidepressants, & anti-anxiety drugs beneficial for some
- Features of autism may develop over time
- Joints may be abnormally flexible
- Heart disease (mitral valve prolapse) may occur

Other Syndromes - Poor Growth

- Smith-Lemli-Opitz (first described 1964)
- Deletion 13q (1963)
- Familial white matter hypoplasia, agenesis of corpus callosum, MR, & growth deficiency (1993)
- Familial Olivopontocerebellar Atrophy (1988)

Neurologic Swallow Deficits

- Spastic quadriplegia - most severe
 - Non-walkers
 - Non-talkers
- CNS based deficits: incoordination
- PNS/neuromuscular deficits: weakness

Principles of Management

- Whole child approach
- Total oral feeding is not always the goal
- Nutrition & respiratory status critical
- GER managed optimally
- Changes in management needed with gains or regression

Management Recommendations

- Direct & indirect approaches for oral sensorimotor function
- Types of abnormal sensory responses need to be considered
- Oral sensorimotor treatment for anatomic structure problems

Intervention Based on Developmental Skill Levels

- Overall gross & fine motor skill levels
- Cognitive, language, communication
- Adjusted age for first year or two in case of prematurity
- Important that all involved with a child understand & respect the child

Ongoing Monitoring for Potential Changes

- Airway status
- GI tract disease (e.g., GER)
- Clinical ongoing assessment
 - Postural/positional observations
 - Caregiver/child interactions
 - Oral sensorimotor feeding status
 - Observation of respiration

Intervention for Dysfunctional Swallowing

- Dietary changes
- Position & posture
- Bolus placement in mouth
- Timing between bolus presentations
- Thermal sensitization - caution for infants & young children

Bolus Formation (Oral-Motor Focus for Function of Structures)

- Jaw
- Lips
- Cheeks
- Tongue
- Palate

Oral Phase Management

- Positioning
- Sensory aspects
- Presentation
- Texture
- Movement patterns

Pharyngeal Phase Management

- Indirect oral sensorimotor treatment (e.g., improve tongue base propulsion)
- Position changes
- Texture changes
- Surface electrical stimulation
 - Data needed
 - Anecdotal reports not adequate

Nutrition Support

- Boost calories in a variety of ways
- Special formulas or foods
- Cut back calories/volume
 - Close monitoring with tube feeds
 - Infants with cardiac conditions along with neurologic problems may be fluid restricted

Positioning & Seating

- Critical as underpinning to oral sensorimotor considerations
- Adaptations may be needed with
 - Hypotonia
 - Hypertonia
 - Growth
 - Regression

Neurologic Problems: Summary

- Airway & nutrition highest priorities
- Often cannot depend on clinic observations alone with suspicion of pharyngeal problem
- Effort expenditure must be considered
 - Developmental skill levels critical
 - Functional techniques/processes

Neurologic Problems: Summary

- Children with neurologically based feeding and swallowing problems are COMPLEX
- Important to understand etiologies & prognosis for direction of change over time
 - VFSS just a brief window in time
- Realistic goals are critical: always keep in mind that children can “beat the odds”