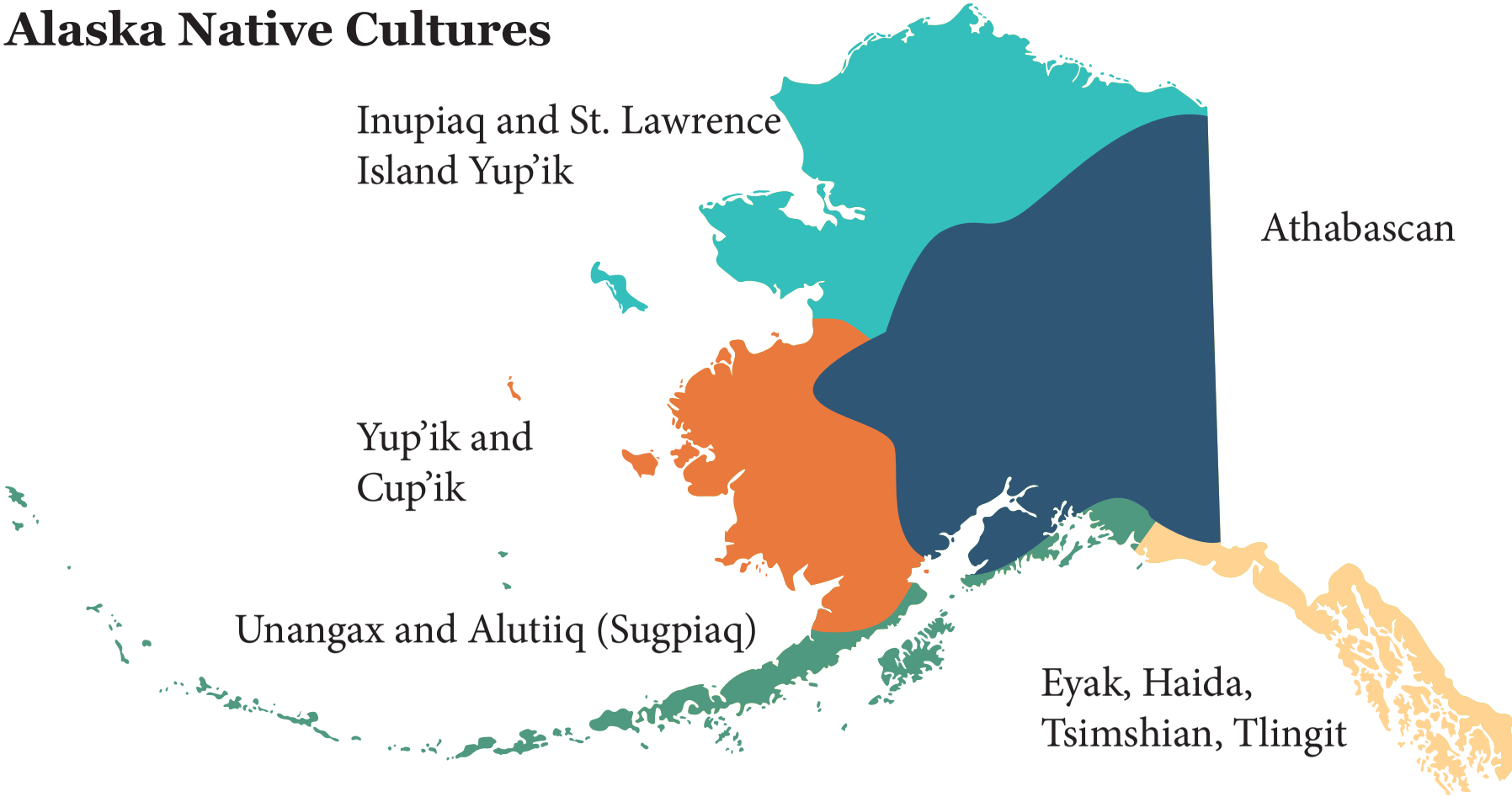
Pocket Guide to Alaska Native Pediatric Diagnoses

Review of diagnoses rarely seen in other populations

ALASKA NATIVE MEDICAL CENTER

VERSION 2: SPRING 2021

Traditional Territories of Alaska Native Cultures



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Pocket Guide to Alaska Native Pediatric Diagnoses:

Review of Diagnoses Rarely Seen in Other Populations

Version 2: Spring 2021

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Directory of Pediatric Specialists and Clinics

For urgent/emergent pediatric questions call ANMC Paging Operator at (907) 563-2662 and ask for the on-call pediatrician.

This is a list of current (as of January 2020) pediatric subspecialties available in Alaska for Alaska Native patients. If you feel your patient needs subspecialty care, please consider referring them to an ANMC Pediatric Regional Field Clinic or ANMC Pediatrics Clinic in Anchorage for further evaluation of subspecialty needs.

Pediatric Subspecialties *Non-ANMC affiliate

- Ped. Cardiology*
- Ped. Dermatology*
- Ped. Endocrinology
- Ped. GI*
- Genetics
  - Visiting from University of Utah/ Primary Children's
- Metabolic Genetics
  - Visiting from Oregon Health and Science University (OHSU)
- Ped. Nephrology*
- Neurodevelopmental Pediatrics
- Neurosurgery
  - General neurosurg based at ANMC
- Ped. Neurology
- Ped. Ophthalmology*
- Ped. Orthopedics
  - General ortho based at ANMC
- Otolaryngology/ENT
  - General ENT based at ANMC
- Ped. Pulmonology*
  - Visiting from Seattle Children's Hosp.
- Ped. Rheumatology
  - Visiting from Seattle Children's Hosp.
- Ped. General Surgery
- Ped. Urology*
- Rehab Medicine
  - Visiting from Seattle Children's Hosp.

Carnitine Palmitoyl Transferase, Type 1A Arctic Variant *(CPT1A Arctic Variant)*

Pathophysiology: Children with two copies (homozygous) of the CPT1A Arctic Variant have a reduced ability to utilize fatty acids from both food and body fat as a source of energy during a prolonged fast.

Demographics:

- The Arctic variant is considered to be the wild-type (normal) form of the CPT1A gene in Inupiaq and Yup'ik populations in Alaska (approx 50% of newborns are homozygous for the Arctic Variant)
- Approximately 7% of all newborns in Alaska are homozygous for the Arctic Variant
- Found at a higher rate in all circumpolar coastline populations including Inuit populations in Canada/Greenland and indigenous populations of northern Siberia
- General population = <1/1,000,000 (CPT1A deficiency due to genetic changes other than the Arctic Variant)

Signs/Symptoms: Most common in children < 2 years old, though most children never have symptoms. Precipitated by a prolonged period of reduced calorie intake (e.g. gastroenteritis), or a prolonged fast.

- Initial signs of metabolic crisis:
  - Sleepiness
  - Irritability
  - Poor appetite
- Metabolic crisis:
  - Hypoketotic hypoglycemia (pathognomonic)
  - Seizures due to hypoglycemia
  - Death, especially associated with a concomitant infectious disease

Identification:

- Alaska Newborn Screen (processed in Iowa) – all children identified by PCR testing beginning July 2016

Management: Avoid fasting states

- Healthy children with CPT1A Arctic Variant should not eat additional calories or more frequently than other children and they can eat the same types of food as other children their age.
- When sick, if infants and toddlers with CPT1A are unable to tolerate glucose-containing fluids (Pedialyte, juice, sports drinks, formula, breast milk) or food for more than 6 to 8 hours, they should see a health care provider immediately for IV, IO, or NG glucose-containing fluids. Oral glucose gel can also be administered.
- Children with CPT1A Arctic Variant who are NPO on IV fluids should always be on dextrose containing fluids (e.g. D5-NS). A normal maintenance rate is all that is needed.
- Prior to surgery, the following guidelines can be followed safely to limit NPO time.
  - NPO 2 hours for clear liquids
  - NPO 4 hours for breast milk
  - NPO 6 hours for formula/non-clear liquids

Critical Times:

Fasting, especially with a concurrent illness or prolonged NPO before surgery, during first 2 years of life

- Fever
- Infection
- Dehydration
- Surgery

For non-urgent questions:

- Matt Hirschfeld, MD - Pediatrician, ANMC
  MHirschfeld@southcentralfoundation.com
- Charlene DiFilippo, RD
  Dietician, SCF Pediatrics
  CDiFilippo@southcentralfoundation.com

For urgent/emergent issues:

- Page the ANMC on-call pediatrician:
  (907) 563-2662

Other resources:

- Newborn Screening Information for Parents: *newbornscreening.info/ Parents/ fattyaciddisorders/CPT1AV.html*
- YouTube - CPT1A AV Videos:
  - Parents: *youtube.com/ watch?v=gE8CnQjZDakandt=639s*
  - Providers: *youtube.com/watch?v= ZBJB6gLAr4Qandfeature=youtu.be*

Congenital Adrenal Hyperplasia (CAH)

Pathophysiology: Deficiency of enzyme required for glucocorticoid and mineralocorticoid production (>90% of cases 21-hydroxylase), resulting in glucocorticoid and mineralocorticoid deficiency and androgen excess.

Cortisol Deficiency →	Hypoglycemia Hypotension
Aldosterone Deficiency →	Low Na+ High K+
Androgen Excess →	Virilization

- Classic Salt Wasting (75%) and Classic Simple Virilizing (25%)
  - Prenatal and progressive postnatal virilization
  - Aldosterone deficiency seen in salt-wasting variant
- Mild, non-classic
  - Mild enzyme deficiency
  - No genital exam abnormalities at birth; variable signs of androgen excess at any phase of postnatal development, can present from birth to teen years

Inheritance: Autosomal recessive

Demographics (by positive NBS):

- Yup'ik = 1:280 live births
- Alaska = 1:4,000 live births
- General = 1:15,000 live births

Signs/Symptoms of initial presentation by age:

- Newborn:
  - Genital exam findings: clitoromegaly and labial fusion (females, classic), scrotal hyperpigmentation (males, classic)
- 1 to 2 weeks old:
  - Adrenal crisis (classic salt-losing): Failure to thrive, dehydration, hyponatremia, hyperkalemia
- 2 to 4 years old:
  - Early virilization with pubic hair,
  - growth spurt, adult body odor (males, classic non-salt-losing)
- School age:
  - Hirsutism, menstrual irregularity, early pubarche, sexual precocity (non-classic, school age children)

Diagnosis:

- Newborn Screen – looks for high levels of 17-OH-progesterone seen in classic CAH

- If NBS(+): check levels of 17-OH-progesterone and electrolytes and consult Peds Endocrinology
- If concerns for non-classic CAH or simple virilizing, call Peds Endocrinology at ANMC
  - ACTH Stimulation Test (measures serum concentrations of 17-OHP before and after giving ACTH)
  - 90-95% sensitive (not necessary for classic CAH)

Management:

- Immediately consult Pediatric Endocrinology (see below) when diagnosed clinically or on the NBS
- If suspicion for CAH, run diagnostic tests and then treat empirically in discussion with Peds Endocrinology. Draw blood for diagnostic tests before treatment. Treat with:
  - Hydrocortisone
  - Fludrocortisone
  - Sodium Chloride
- Adrenal Crisis management (discuss with Endocrinology or ANMC Pediatric Hospitalist on-call in an emergency)
  - Fluids (20mL/kg NS then D5NS or D10NS maintenance)
  - Monitor glucose and electrolytes

- Stress-dose: Hydrocortisone (IV or IM)
- <3yo: 25mg bolus followed by 50mg/m2/day divided q6-8hrs
- 3-12yo: 50mg bolus followed by 50mg/m2/day divided q6-8hrs
- >12yo: 100mg bolus followed by 50mg/m2/day divided q6-8hrs

Critical Times for Affected Patients:

Any time that could trigger adrenal crisis (hypotension, hyponatremia, +/- hyperkalemia, metabolic acidosis, hypoglycemia)

- First 1 to 4 weeks of life (if undiagnosed)
- When ill or severely stressed (e.g. infectious diseases, surgical procedures, trauma etc)

For non-urgent questions:

- Rachel Lescher, MD
  Pediatric Endocrinologist, ANMC
  (907) 563-2662
- Agnes Hunt - Admin
  (907) 729-8822
- Sherry Hammock - Case Manager
  (907) 729-8803
- Paging operator: (907) 563-2662
  Ask for the on-call ped. endocrinologist

Congenital Sucrase-Isomaltase Deficiency (CSID)

Pathophysiology: Lack intestinal brush border enzyme to breakdown di- and oligosaccharides including sucrose and isomaltose

Inheritance: Autosomal recessive (potential mild form in carriers)

Demographics:

- Likely 3-10% of Alaska Native people (exact numbers not known)
- Higher prevalence seen along the western and northern coasts, primarily among the Yup'ik and Inupiaq

Signs/Symptoms:

- Watery diarrhea with sucrose-containing food
- Malnutrition
- Abdominal pain/ distension
- Faltering growth

Diagnosis:

- Genetic screen (blood), run at: Fulgent Genetics *fulgentgenetics.com/congenital-sucrase-isomaltase-deficiency*
- Known mutation ($200) and single gene testing ($895) available (institutional billing cost, as of fall 2020)

Management:

- Breast milk and most formulas are lactose containing (e.g. Similac advance and Enfamil Enfacare) are tolerated
- Dietary Modification: Avoid sucrose, isomaltose and maltose (corn syrup is sucrose).
- Enzyme Replacement: Sucraid (sucrose digestion only), costs~$2,000/month, not covered by Alaska Medicaid at this time, usually covered by most private insurance
- Refer patients needing Sucraid to the Tribally-Sponsored Health Insurance Program (T-SHIP) for eligibility screening and enrollment assistance
  - T-SHIP contact info:
    (907) 729-5696, 1(888) 770-7346
    anthctship@anthc.org.

Critical Times for Affected Patients: When first exposed to sucrose (some formulas; most often when starting solids around 6mo old). Consider this diagnosis when you have an infant or toddler with chronic diarrhea who has recently started solids or transitioned from breast milk or formula to whole milk and other foods.

For further questions:

- Matt Hirschfeld, MD - Pediatrician, ANMC
  MHirschfeld@southcentralfoundation.com
- Charlene DiFilippo, RD
  Dietician, SCF Pediatrics
  CDiFilippo@southcentralfoundation.com
- Sam Maloney, RD - Dietician, ANMC
  SMaloney@anthc.org

Other resources:

- *csidcares.org*
- *anthc.org/csid*
- ANMC CSID diagnosis and treatment guidelines: *anmc.org/files/CSID.pdf*

CSID Dietary Recommendations:

Each patient with CSID responds to foods variably. If a patient tolerates a food listed below without diarrhea, then there is no reason to limit intake.

Fruits to avoid	Apples, apricots, bananas, cantaloupe, dates, grapefruit, guava, honeydew, mango, nectarine, oranges, tangerines, passion fruit, peaches, pineapple, tangerines
Fruits typically tolerated	Avocado, blackberries, blueberries, boysenberries, cherries, cranberries, fresh currants, grapes, kiwifruit, lemons, limes, olives, papaya, pears, pomegranates, prunes, raspberries, rhubarb, strawberries
Vegetables to avoid	Beets, black beans, black-eyed peas, butternut squash, carrots, cassava (yuca), carrots, chickpeas, corn, garlic, green peas, lentils, kidney beans, lima beans, navy beans, onions, parsnips, pinto beans, potatoes
Vegetables typically tolerated	Collard greens, cress, cucumber, eggplant, endive, green beans, kale, lettuce, mung bean, sprouts, mushrooms, mustard greens, okra, peppers, radishes, rutabaga, snow peas, turnips, yellow squash, zucchini

Kuskokwim Syndrome (Arthrogryposis-like syndrome)

Pathophysiology: Mutation in the FKBP10 gene resulting in impaired collagen cross-linking and disorganization of collagen molecules causing congenital joint contractures

Inheritance: Autosomal recessive

Demographics:

- Rare, incidence unknown
- Found only in Yup'ik population

Signs/Symptoms:

- Contractures are generally present at birth, worsen during childhood, then stabilize
- Range and severity of contractures varies greatly
- Often contractures of large joints, especially knees and elbows with other lower extremity joints often involved

- Milder skeletal features are common including:
  - Spine: scoliosis, lordosis, spondylolisthesis
  - Feet: bunions (hallux valgus), flat feet (plano valgus), club feet (talipes equinovarus)

Diagnosis:

Genetic testing – discuss with ANMC On-Call Pediatrician and ANMC Orthopedics

Management:

- Bracing and surgical correction of lower extremity contractures to allow ambulation
- OT and PT to enhance upper limb movement for self-care and lower limb movement for ambulation
- Consider bisphosphonate use in consultation with pediatric orthopedics and endocrinology

For non-urgent questions:

- ANMC Orthopedics
  ANMC Paging Operator: (907) 563-2662

For urgent/emergent issues:

- Page the ANMC on-call pediatrician:
  (907) 563-2662

Other resources:

- NIH Genetic and Rare Diseases Information Center: *rarediseases.info.nih.gov/gard/3150/kuskokwim-disease/resources/1*
- National Library of Medicine Genetics Home Reference: *ghr.nlm.nih.gov/condition/kuskokwim-syndrome*

Metachromatic Leukodystrophy (MLD)

Pathophysiology: Lysosomal storage disease causing progressive demyelination of central and peripheral nervous system, also affecting kidneys and other visceral organs due to accumulation of cerebroside sulfate

Inheritance: Autosomal recessive

Demographics:

- 1:2,500 in Navajo (related to Athabascan)
- 1:40,000-1:100,000 in northern Europe and North America

Signs/Symptoms: Children have normal development until disease onset

Type	Age	Symptoms
Late infantile onset = more common AK variant	6 mo-4 yrs	Regression of motor skills, gait difficulties, hypotonia, seizures, ataxia, hypotonia, extensor plantar responses, optic atrophy, fussiness and pain(thought to be due to neuropathy or dystonia)
Juvenile and adult onset	>4 years	Gait disturbance, ataxia, seizures, intellectual impairment, behavioral difficulties, upper motor neuron signs, peripheral neuropathy

Diagnosis:

- Brain MRI
  - symmetric white matter lesions with periventricular predominance (early) and cortical atrophy (late)
- Genetic testing with leukodystrophy panels from GeneDx or Invitae. Details for sample requirements available online.
- Previously, testing was performed for ARSA (arylsulfatase A) gene activity, but genetic panels allow for testing for a wider range of genes and mutations.

Management:

- Requires multidisciplinary management: Refer to Neurology, GI, Nutrition, PT, OT, SW
- No curative treatment – prognosis for late infantile and early juvenile onset is poor (death within 5-6 years)
- Bone marrow transplant, gene therapy and hematopoietic stem cell transplant are all investigational with goal of slowing the disease course. Effectiveness of transplant requires diagnosis prior to onset of symptoms.
- Discuss with genetic counseling regarding testing for subsequent children

For non-urgent questions:

- Providence Pediatric Neurosciences
  (907) 212-2321

For urgent/emergent issues:

- Page the ANMC on-call pediatrician
  (907) 563-2662

Other resources:

- National Library of Medicine Genetics Home Reference: *ghr.nlm.nih.gov/condition/metachromatic-leukodystrophy*
- GeneDx panel: *genedx.com/test-catalog/available-tests/leukodystrophy-xpanded-panel*
- Invitae panel: *invitae.com/en/physician/tests/06174/#info-panel-disorders_tested*

Micro-aspiration in Apparently Neurologically Typical Children

Pathophysiology: Unknown

Demographics: Unknown, but appears to be a significant number of otherwise normal children under age 3 from western and northern Alaska

Signs/Symptoms:

Overt signs/symptoms not always present

- Frequent cough
- Aspiration with feeds during URIs
- Frequent (or any) pneumonia, especially right upper lobe

Diagnosis:

Clinical history/bedside evaluation vs Video Fluoroscopic Swallow Evaluation (VFSS) Fiberoptic Endoscopic Evaluations of Swallowing (FEES)

- Radiation exposure from video fluoroscopic swallowing study plus inconvenience of transporting families to Anchorage can be avoided by initially treating symptomatic patients
- Limited equipment/staff to perform VFSS/FEES in rural Alaska
- During a global pandemic, FEES may be problematic due to possible aerosol and droplet generation due to cough, gag or sneeze

Management:

- Slowing nipple flow rate at breast or bottle (e.g. layback breast feeding position or decrease flow to a Dr. Brown's Preemie or transition nipple). Ensure that families do not cut or alter the nipples in any way
- Infant driven feeding practice which aims to monitor infant communication cues and provide supports (pacing, positioning, etc.) before the child becomes fatigued and/or has an aspiration occurrence

- Speech Language Pathologists at ANMC are available for training in reducing flow rate and/or infant driven feeding practices.
- Thickening milk to "slightly thick" or "mildly thick" as a strategy to reduce flow rate and increase bolus cohesion. If successful, continue thickened liquids until the patient experiences 12 symptom-free months, then gradually wean off thickener. If child does not tolerate wean, refer to ANMC for swallow evaluation.
- Management strategies above require close follow-up to document improvement vs no change vs worsening lung disease. If still experiencing overt symptoms of aspiration or if no resolution of lung disease within 3 months, child should be referred to ANMC for a video fluoroscopic swallow study/modified barium swallow study.
  - **Thickening agents:** Thick-it, Simply Thick, rice/oatmeal baby cereal. Thick-it and infant cereal can only be used and mixed to mildly thick (IDDSI level 2). Recipes differ according to thickener agent and formula type. Contact speech therapy at ANMC (contact numbers below) for further instruction.
  - There is risk that a child may still be aspirating with thickener, but with

reduced signs and symptoms, leading to potential for aspiration of thickener. Clearance can be more difficult via spontaneous cough due to thickened consistency. As noted, close follow up is critical if treating empirically.

Critical Times for Affected Patients:

Respiratory illness in infancy, usually less than 3 years old

For questions:

- Matt Hirschfeld, MD - Pediatrician, ANMC
  MHirschfeld@southcentralfoundation.com
- ANMC Outpatient Speech Therapy
  (907) 729-6632
- ANMC Inpatient Speech Therapy
  (907) 729-1063

Other resources:

- Duncan et al. Curr Gastroenterol Rep. 2019 May 16;21(7):30.
- *pubmed.ncbi.nlm.nih.gov/31098722*
- Rempel et al. Pediatr Pulmonol. 2006 Oct;41(10):912-5.
- *pubmed.ncbi.nlm.nih.gov/16933218*
- Rempel et al. Pediatr Pulmonol. 2011 Dec;46(12):1240-6. *pubmed.ncbi.nlm.nih.gov/21618720/*
- *iddsi.org*

Optic Nerve Hypoplasia (ONH)

With or without hypopituitarism; also known as Septo-Optic Dysplasia (SOD) and de Morsier Syndrome

Pathophysiology: Disorder of early brain development resulting in spectrum of findings including hypoplasia of optic nerve, agenesis of corpus callosum and septum pellucidum, and/or pituitary hypoplasia

Inheritance: Usually sporadic; occasionally autosomal recessive

Demographics:

- 1:10,000 live births
- Unknown but anecdotally higher incidence for Alaskan Native

Signs/Symptoms:

- Hypoplasia of optic nerve
  - impaired vision (one or both eyes)
  - nystagmus

- Abnormal midline brain structure formation (corpus callosum, septum pellucidum)
  - intellectual disability
  - other neurologic problems including seizures
- Pituitary anomalies (hypoplasia, ectopia, etc.)
  - Growth hormone deficiency (most common)
  - Various pituitary hormone insufficiencies
  - Pituitary insufficiencies may evolve
- Cleft palate

Diagnosis:

- Brain and Pituitary MRI with and without contrast
  - Thinning of optic nerves and chiasm
  - Absence of septum pellucidum
  - Agenesis or hypoplasia of the corpus callosum
  - Pituitary hypoplasia or posterior pituitary ectopia
- Ophthalmology exam
- Endocrinology evaluation
- Can be suspected initially based on prenatal ultrasound

Management: Varies depending on individual

- Refer to Pediatric Endocrinology for regular endocrine evaluations

- Refer to Ophthalmology
- Refer to Infant Learning Program/Birth to 3
- Refer to Pediatric Neurology in setting of seizures and neurologic deficits

Critical Times for Affected Patients:

Vary depending on individual

- If hypopituitarism, times of stress (fasting, illness, surgery, trauma) are high risk as well as newborn period due to:
  - ACTH/Cortisol deficiency can present with adrenal crisis in the first week of life (similar to CAH). This is NOT picked up on the newborn screen.
  - GH and/or ACTH deficiency can present with hypoglycemia
  - Thyroid deficiency if poor eating or fatigue (will not be diagnosed on AK newborn screen)
  - Diabetes insipidus can present in early infancy with poor weight gain or weight loss, polyuria, polydipsia, dehydration, hypernatremia

For non-urgent questions:

- Rachel Lescher, MD
  Pediatric Endocrinologist, ANMC
  (907) 729-8822
- Agnes Hunt - Admin
  (907) 729-8822

- Sherry Hammock - Case Manager
  (907) 729-8803
- Kevin Winkle, MD
  Pediatric Ophthalmologist
  (907) 561-1917, rkwinkle@anthc.org
- Providence Pediatric Neurosciences
  (907) 212-2321
- Contact ANMC Pediatric Hospitalist via ANMC Paging Operator: (907) 563-2662

For urgent questions:

- Page the ANMC on-call pediatrician: (907) 563-2662

Other resources:

- National Library of Medicine Genetics Home Reference: *rarediseases.info.nih.gov/diseases/7627/septo-optic-dysplasia-spectrum*

Non-CF Bronchiectasis

Pathophysiology: Recurrent lower respiratory tract infections (especially pneumonia) causes airway inflammation and damage that leads to airway dilation and reduced elasticity as well as loss of muco-ciliary function with difficulty clearing secretions. Most common airway organisms include H Flu (non-typeable), Strep pneumo, Moraxella catarrhalis.

Risk factors: Recurrent pneumonia/ bronchiolitis in first year of life, prematurity, household crowding, lack of running water, indoor air pollutants

Demographics:

- More common among indigenous children globally (AK, New Zealand, Australia)
- Alaska Yukon-Kuskokwim Delta: 1 in 63 children
- U.S.: 1 in 250,000.

Signs/Symptoms:

- Chronic wet cough
- Recurrent respiratory exacerbations with fever, crackles/wheezing with persistent infiltrate on chest X-ray

Diagnosis: There is a clinical continuum beginning with protracted bacterial bronchitis (PBB) to chronic suppurative lung disease (CSLD) to bronchiectasis (irreversible)

- PBB - isolated wet cough >4 weeks without underlying disease or specific cough pointer
- CSLD - 3 episodes of wet cough lasting >4 weeks over one year
- Bronchiectasis - clinical features of CSLD with radiographic findings of bronchiectasis (dilation, thickened airway walls, and saccular changes)

High resolution CT (HRCT) is used to confirm the diagnosis of bronchiectasis.

Treatment:

Refer to the Yukon-Kuskokwim Health Corporation (YKHC) algorithm below

- Acute management:
  - Antibiotics (Augmentin 45 mg/kg/dose BID or Cefidinir 14 mg/kg/dose)x2-4 wks
  - Airway clearance x2-4 wks
- Chronic management:
  - Refer to pulmonology (Anchorage/ Bethel). Yearly PFTs after age 5
  - Assess for and manage co-morbidities (aspiration, asthma, TB) and ensure vaccinations appropriate (PCV-13 plus PPSV-23 x 1 dose after age 2)
  - Optimize environmental health, nutritional status, and airway clearance

Critical times in management:

- Early identification and management prevents long-term sequelae (decline in lung function and early COPD)
- Exacerbations - treat aggressively

For non-urgent questions:

- Pediatric Pulmonology at Seattle Children's Hospital
  - Quarterly visits to ANMC and YKHC
- SCH provider line: (206) 987-7777 Request pulmonology

For urgent/emergent issues:

- Page the ANMC on-call pediatrician: (907) 563-2662

Resources:

- YKHC Clinical Guideline for Bronchiectasis
- Chang AB et al. Management of Children With Chronic Wet Cough and Protracted Bacterial Bronchitis: CHEST Guideline and Expert Panel Report. Chest. 2017 Apr;151(4):884-890. doi: 10.1016/j.chest.2017.01.025. Epub 2017 Jan 28.
- Kinghorn B, Singleton R, Mccallum GB, Bulkow L, Grimwood K, Hermann L, Chang AB, Redding G. Clinical course of chronic suppurative lung disease and bronchiectasis in Alaska Native children. Pediatric Pulmonology 2018;1-8.

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