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THE PEDIATRIC JOURNEY

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CLEFT LIP AND PALATE - WHAT TO DO?

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Kids with cleft lip and palate present a significant challenge to any provider involved in their care. Cleft lip and palate is the most common facial deformity (1:700 live births). Our region is particularly vulnerable due to a couple of prevalent factors: poor prenatal care leading Latino and Native American populations which are statistically more vulnerable to cleft lip and palate deformities.

SUSPECTED CULPRITS The etiology of cleft lip and palate is mutifactorial and has been associated with genetic factors, teratogenic drugs, nutritional deficiencies, amniotic banding, chemical exposures, maternal hypoxia, and radiation. Most clefts in familes are sporadic, however, if 1 parent has a cleft there is a 4-6% chance that their baby will also have a cleft. Interestingly, boys are twice as likely to have a cleft lip (with or without palatal involvement) and girls are more likely to have an isolated cleft palate only.

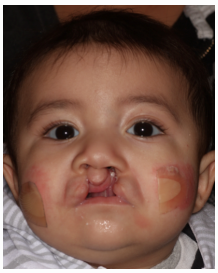
ARE THEY SYNDROMIC? If an isolated cleft palate is present there is approximately a 50% chance that the baby will have an associated syndrome. This is in contrast to a cleft lip patient (with or without palatal involvement) which is only likely to have an associated syndrome in approximately 10% of the cases. **In patients with an isolated cleft palate there should be a lower threshold for a thorough work-up if the patient is not progressing as expected.**

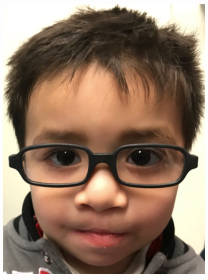
WHEN DO THEY GET THEIR SURGERIES (TABLE 1) The timing of cleft surgery historically has been extremely controversial. However, of late, national recommendations based on research have helped to standardize cleft care. If one operates too early the patient will suffer **growth restrictions** that will likely require additional future surgeries to correct. However, early surgery may still be indicated secondary to **functional or psychosocial** concers.

TABLE 1 - WHEN TO DO SURGERY?

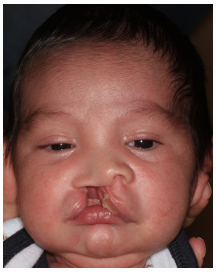
Surgery Needed	Recommended Time for Repair	Rationale
Cleft Lip Surgery	3-5 months of age	1) Safer anesthetic than when newborn 2) Completion of nasoalveolar molding
Cleft Palate Surgery	10-18 months of age	Speech development
Cleft Maxillary Bone Graft	6-12 years of age	Essential to perform before adult teeth erupt into cleft site
Cleft Jaw Surgery (Orthognathic Surgery)	15-20 years of age	50% of cleft patients need this surgery to correct jaw discrepancies
Cleft Nasal Surgery (Rhinoplasty)	15-16 years of age	1) Nasal obstruction 2) Nasal deformity
Speech Surgery	6-12 years of age	30% need additional surgery to aid in speech outcomes
Cleft Lip Revision	5-18 years of age	Nationally every cleft lip is revised about 2.1 x during the patients life

BILATERAL CLEFT LIP AND PALATE





UNLATERAL CLEFT LIP AND PALATE



WHAT DOES THE CLEFT & CRANIOFACIAL TEAM LOOK FOR?

Cleft care is complicated - these kids are likely to fall through the cracks and all too often may only have limited exposure to an **American Cleft Palate - Craniofacial Association (ACPA)** approved cleft and craniofacial Team. The primary care physician, school

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nurse, or speech therapist may be their only contact with a healthcare professional. It is also assumed by many that "others" are monitoring their complicated facial issues and "of course" they must be plugged into a Cleft or Craniofacial Team - but all too often this is not the reality. When seeing ANY cleft patient it is critical to keep in mind what problems they are likely to be experiencing.

- Does their speech sound appropriate? Are they having difficulties in school?
- Are they growing their hair out to cover their face?
- Are their teeth appropriate or do they suffer from dental disease?
- Are they experiencing sleep apnea?
- Are they socially isolated or thriving in their school environment?
- Do they still have an obvious deformity? Do liquids or food come out of their nose when they eat? **(TABLE 2)**

WHAT CAN WE DO?

It is CRITICAL that these kids are involved in an ACPA apprioved Cleft and Craniofacial Team. Our Cleft and Craniofacial Team is composed of individuals who have dedicated their practices specifically to Cleft and/or Craniofacial care. Our El Paso ACPA approved team includes Cleft and Craniofacial Surgeons, Ear Nose and Throat Surgeons, Oral and Maxillofacial Surgeons, Neurosurgeons, Craniofacial Orthodontist, Pediatric Dentists specializing in Cleft/Craniofacial deformities, Speech and Language Pathologist, Occupational Therapist, Nutritionist/Dieticians, Nursing Coordinators, and Social Workers.

The Primary care physician is the gatekeeper in the fight to ensure success for kids with cleft lip and palate, but truly all of us are responsible once we come in contact with an affected child. These kids are at risk, and it is essential that vigilance is maintained. Hard and probing questions need to be asked to the child and parents concerning the child's overall welfare. Treating this population has been one of the highlights of my career - these kids are resourceful, intelligent, fun, and resilient. I have and continue to learn many life lessons from them.

TABLE 2 - PROBLEMS ASSOCIATED WITH CLEFTS

Contact:
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El Paso Cranial and Facial Clinic - 915-242-8500
High Desert Oral and Facial Surgery Clinic - 915-833-2969

THE HEAT IS ON - UNDERSTANDING HEAT ILLNESS AND MANAGING HEAT STROKE

BY: ARJUN CHANDRAN - PICU INTENSIVIST

Heat -related illness represents a continuum of disease states. When the heat index is higher than 95F (35C), mortality increases in relation to elevation of temperature and duration of the heat wave. In most cases, these fatalities are preventable. Children, people with heart and lung diseases, those with chronic mental disorders and those taking medications that interfere with salt and water balance (diuretics, antihypertensives etc) are also at increased risk. Early recognition of symptoms and rapid cooling are crucial, because heat stroke is a medical emergency.

THERMOREGULATION AND HEAT ACCLIMATIZATION

Thermoregulation occurs through a series of voluntary (cerebral cortex) and involuntary mechanisms (hypothalamic). Peripheral vasodilation occurs to augment dermal blood flow, which allows convective heat loss into the surrounding air. In an effort to perserve central perfusion, there is vasoconstriction of the peripheral vasculature creating hypoperfusion. When a person's core temperature is 104 F or greater, cellular damage occurs. There is a systemic inflammatory response, and increased cell wall permeability allows the release of endotoxins. These disruptions of homeostasis initiate a cascade of events that include tissue hypoxis, metabolic acidosis, and servere organ dysfunction.

Acclimatization are adaptions that allow us to tolerate heat. These can take weeks to develop: Enhancement of cardiovascular performance, activation of the renin-angiotensin-aldosterone axis, salt convservation by the sweat glands and kidneys, increase in the capacity to secrete sweat, expansion of plasma

volume, increase in the glomerular filtration rate, increase in the ability to resist exertional rhabdomyolysis.

Children are prone to heat-related illness because of their greater surface area to body mass ratio. This permits more heat transfer from the environment. They also have diminished ability to dissipate heat because of slower sweat rates, a higher temperature threshold for the initiation of sweating, and production of a more diluted sweat. Furthermore, they are slower to acclimatize to the heat and have less of a thirst response.

SPECTRUM OF HEAT ILLNESSES:

HEAT EDEMA:Benign, self-limiting. Lower extremities, secondary to prolonged cutaneous vasodilation

HEAT RASH (LICHEN TROPICS, PRICKLY HEAT, MILARIA RUBRA): Plugging of sweat ducts, inflammatory eruption.

HEAT CRAMPS:Exercise - associated muscle contractions, believed to be secondary to electrolyte losses.

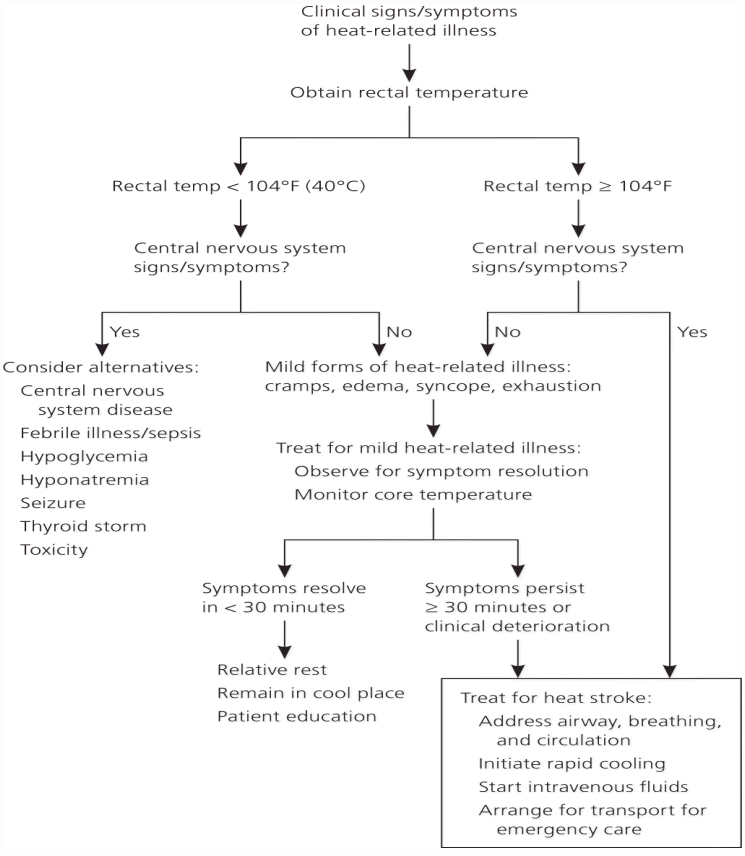
HEAT TENTANY:Carpopedal spam and paresthesia secondary to hyperventilation.

HEAT SYNCOPE:Transient loss of consciousness or collapse, rapid return to baseline.

HEAT EXHAUSTION:Loss of water and salt following exposure with fatigue, rapid pulse, profuse sweating, vomiting and weakness. NO CNS INVOLVEMENT

HEAT STROKE (MOST SEVERE): TWO TYPES: EXPOSURE AND EXERTIONAL (EHS): Core Temp >40, CNS involvement with delirium, convulsions or coma.e

EVALUATION OF A HEAT ILLNESS:



HEAT STROKE AND ITS MANAGEMENT:

Heat stroke is the most servere form of heat illness. It is further classified as 'Classical' when it occurs secondary to exposure to high environment temperature or 'Exertional' when seen in previously healthy young poeple, exercising in hot and humid climate.

Hyperthermia and central nervous system dysfunction must be present for a diagnosis of heat stroke. The core temperature may range from 40C to 47C.

Brain dysfunction is usually servere but may be subtle, manifesting only as inappropriate behavior or impaired judgment; more often, however, patients have delirium or frank coma. Seizures may occur, especially during cooling.

Hypophosphatemia and hypokalemia are common at the time of admission. Hypoglycemia is rare. Hypercalcemia and hyperproteinemia, reflecting hemoconcentration, may also occur. The most serious complications of heat stroke are those falling within the category of multiorgan-dysfunction syndrome. They include encephalopathy, rhabdomyolysis, acute renal failure, acute respiratory distress syndrome, myocardial injury, hepatocellular injury, intentinal ischemia or infarction, pancreatic injury, and hemorrhagic complications, espcially disseminated intravascular coagulation, with pronounced thrombocytopenia.

Cooling measures need to initiate to bring core body tempurature below 39.4 C, within 30 minutues of presentation. There are serveral methods of cooling:

CONDUCTIVE COOLING: A. Ice-water immersion
B.Application ice packs

EVAPORATION: Continuous spraying of water over the skin combined with foced-air equipment (ventilator/fan)

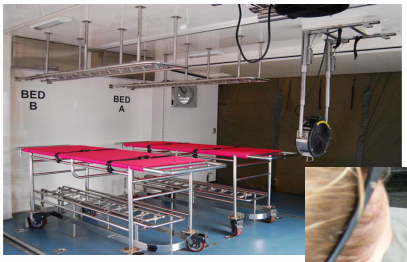
INVASIVE COOLING: A. Iced peritoneal lavage.
B. Iced gastric lavage.
C. Intravascular cooling catheter, being studied in EHS

COLD IVF: Adjunctive, insufficient as primary treatment

ALCOHOL SPONGE BATHS: S/E: Systemic absorption

COOLING BLANKETS: Not well studied, unproven efficacy

CRRT/PLASMAPHERESIS: For cooling and removal of inflammatory cytokines. Useful in multi-organ dysfunction/organ failure by supportive management.



CONCLUSION

Rapid access to cooling is the most effective method to prevent heat stroke fatalities. Athletic events should be held during cooler periods of the day and scheduled to avoid hot, humid months. Athletes, coaches and parents should be edcucated about the early signs of heat-related illness, hydration, and initial treatments.

Heat acclimatization is the best protection against heat-related illness-a gradual progression of duration and intensity of exercise over 10 to 14 days. Proper hydration, lightweight and light -colored clothing, shaded rest areas, and rule adjustments to allow for frequent substitutions can also help protect young athletes. Automobiles should be locked, and children should never be left unattended in an automobile during hot weather. Deaths during heat waves are still common and have associated largely with social isolation in vulnerable populations, lack of air conditioning, and increases in heat during large gatherings for cultural or religious purposes. Encouraging the use of weather apps, employing city and state resources to alert authorities and individuals taking care of at risk persons, providing air conditioned shelters-especially in border cities, and reducing energy costs during weather so that air conditioning is affordable may decrease morbidity and mortality during heat waves. heat stroke. The core temperature may range from 40C to 47C.

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2019 PEDIATRIC GRAND ROUNDS

The First & Third Wednesday of Every Month

Breakfast: 7:30-8 a.m. Grand Rounds: 8-9 a.m.
Academic Education Center (AEC), 2nd Floor, 4800 Alberta Avenue

SEPTEMBER 18TH, 2019	Mechine Learning and Big Data: Real Time Quality Improvement in Healthcare	Punkaj Gupta, M.D.
OCTOBER 2ND, 2019	Cytogenetic Testing and Findings for Neonates and Constitutional Disorders in Pediatric	Daniel Bustamante
NOVEMBER 6TH, 2019	Asthma in the ICU	Arjun Chandran, MD
DECEMBER 18TH, 2019	NO GRAND ROUNDS	NO GRAND ROUNDS

SATURDAY, OCTOBER 5TH

8AM REGISTRATION
9AM GOLF START