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Pre and Perioperative considerations in a patient with Cold Induced Anaphylaxis and recently diagnosed Hereditary Angioedema

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Summary
Severe cold urticaria and hereditary angioedema can cause immediate, life threatening respiratory compromise from increased vascular permeability of the oropharynx. Pre and perioperative evaluation of patients with a history of these conditions must be geared toward eliminating known triggers, prophylactic treatment with plasma-derived C1-inhibitor, and immediate intervention in an acute episode.

Case Report
We describe a 14 year old female presenting for the removal of a knee mass acquired after a trauma. The patient had a recent ED visit for acute onset periorbital swelling, wheezing and difficulty breathing requiring administration of epinephrine for symptoms of anaphylaxis. Prior to surgery, the patient was diagnosed with Hereditary Angioedema after an outpatient specialist found low normal levels of C1 esterase inhibitor and a low level of C4 complement. Her history was significant for anaphylaxis as well as intolerance to cold. The patient was pretreated with Cinryze, human plasma-derived C1-inhibitor, two days, and then again, one hour prior to her intended procedure. In addition, the patient was scheduled to receive two additional doses up to one week post-operatively. During preoperative preparation, the patient was pre-treated with forced air warming blankets. The operating room was warmed to 25°C. On arrival to the operating room, routine ASA monitors were placed. Upper and partial lower body forced air warming blankets (set at 41°C) were placed on non-surgical field areas as well as underbody heating. A fluid warmer (set at 41°C) was used for all intravenously administered medications and fluids. Pre-oxygenation with 100% oxygen with an HME commenced for two minutes prior to IV induction with 150mg of Propofol and 100mcg of Fentanyl. A size 3 laryngeal mask airway was placed without difficulty. Anesthesia was maintained with SevoFlurane. The intraoperative course continued without event. The patient received IV acetaminophen, ondansetron, and dexmedetomidine for prevention of emergence delirium prior to extubation. Upon completion of the procedure, she was gently suctioned and LMA was removed without event. She was transferred to PACU with oxygen via nasal cannula and pulse oximetry and heated blankets. The post-operative course was uneventful. The patient’s vital signs remained stable, with oral temperature readings measuring between 37.1°C to 37.7°C. Ultimately, the patient was discharged from PACU to home in stable condition with no concerns for recovery.

Discussion
Cold urticaria is classified as a type I (immediate) hypersensitivity syndrome which can cause systemic disease with cutaneous manifestations. The most common cutaneous manifestations are transient, well circumscribed edematous skin lesions that are erythematous and pruritic. In some cases, wheals can occur in response to physical stimuli such as exposure to cold air or water. In susceptible patients, immersion in water may lead to respiratory and cardiovascular compromise, syncope, and, ultimately, drowning. Patients with oropharyngeal reactions to cool liquids and other food and children are particularly at risk for the development of systemic and anaphylactic reactions. Relative hypothermia is a well-documented complication of general anesthesia. Patients undergoing general anesthesia experience a rapid decrease in core temperature in the first hour after induction due to the redistribution of heat from the core to the periphery. Additional causes of intraoperative hypothermia include neonatal population, low OR temperature, burn patients, and procedures complicated by high amounts of blood loss.

Approximately 35-40% of patients with acute or chronic urticaria can also experience angioedema. Angioedema can be seen as a component of urticaria or as a distinct entity. It is characterized by swelling of the lips, hands, gastrointestinal tract, or in rare cases the tongue and oropharynx leading to life-threatening airway obstruction. Hereditary angioedema (HAE) is a rare (1:10,000 – 1:150,000), autosomal dominant disorder caused by deficiency of C1-inhibitor resulting in the uncontrolled release of bradykinin, eventually leading to an increase in vascular permeability and fluid leakage, resulting in subcutaneous and mucosal edema. Acute attacks often last 72-96 hours, and are often severe with over half of patients suffering at least one upper airway attack in their lifetime.

Acute attacks of HAE can be triggered by minor trauma, emotional stress, infection, cold exposure, certain food or drugs. Surgical trauma, airway manipulation, even dental procedures have been noted to cause fatal attacks. The tentative nature of the disease, uncertain triggering stimuli, and the possibility of potentially fatal repercussions, especially in the setting of surgical and airway manipulation, makes HAE of particular concern to anesthesiologists. The concern is especially great in the pediatric population since patients have a much narrower airway and estimates of mortality from an acute laryngeal attack near 15-33%. Formal recommendations for pre and perioperative management of patients with HAE are limited due to the scarcity of the disease. From the anesthesiologist’s perspective, eliminating any known trigger is essential. Pre-operatively, short term prophylaxis with human plasma derived C1-inhibitor should be administered one hour prior to procedure, and should be immediately available post-operatively. C1-inhibitor levels should be kept optimally therapeutic prior to procedure and should be assessed prior to the day of surgery. Some reports show utility in giving C1-inhibitor peri-operatively. In addition, limiting airway manipulation and inhibiting the stress response by implementing regional anesthetic techniques would likely be preferable. When regional anesthesia is not feasible, commonly implemented modalities of general anesthetic (volatile agents, IV anesthetics, NMBA) have no known contraindications. Intra-operatively, extra precautions and equipment should be readily available for management of difficult airways. Post-operative monitoring is also essential as the time course of an acute attack can be variable.

Severe cold urticaria and hereditary angioedema can be fatal in times of stress and, particularly, with surgical stimulation and airway manipulation. Pre and perioperative management of these patients requires formal guidelines for prophylactic therapy, induction and airway guidelines, use of advanced airways and post-operative monitoring and surveillance. Conservative options should be
preferred when available; however, use of best practices must be established for patients requiring surgical intervention.