HISTORY: Elizabeth Milo is known to have dysautonomia, mast cell activation syndrome (MCAS), a connective tissue disorder (EDS), and poor GI motility. Consequently, there are several issues that need to be addressed to make the surgical experience better.

The most important considerations in emergency situations and for surgery are:

- My reactivity and hyper-sensitivity to medications, which necessitates very low dose and slow administration (*see attached allergy list*).
- I HAVE NEVER HAD ANESTHESIA and cannot predict how I will respond to medications. If possible, start with Propofol *only* and at lowest dose possible.
- Maintaining my low blood pressure and low blood sugar.
- My history of idiopathic and atypical anaphylaxis with syncope (*see attached anaphylaxis protocol*).
- Avoiding vasodilators and histamine-releasing agents (*see attached medication guideline*).
- I have craniocervical instability: Use Glidescope or fiberoptic intubation and the most conservative c-spine protocols reserved for patients with neck injuries.

BEFORE SURGERY DISCUSSIONS TOPICS:

- Before surgery, consider trialing any medications that patient has never had (anesthetic agents, D5, LR, Tramadol, benzos etc). Also consider pre-surgery tests, such as lung vital capacity, 24-hour urine free cortisol, serum cortisol, liver panel, RBC magnesium, serum potassium and other electrolytes.
- Medication and supplements (*see attached list*). Stop non-essential medications and supplements at least a week before surgery.
- Premedications (H1/ H2-blockers, corticosteroids etc) and whether I need increased dose of my daily hydrocortisone.
- Anesthetic medications. I am unusually reactive to small amounts of medications. All medications should only be used if absolutely necessary (for example, prophylactic nausea meds and local anesthetics are probably not needed) and then used sparingly and in small doses until my response can be assessed. ***See attached mast cell medication guidelines***
- Oxygen use throughout procedure and inform anesthesiologist about sleep apnea and asthma.
- Anaphylaxis protocol: H1 and H2 antihistamines, corticosteroids, oxygen, IV fluids, bronchodilators. Emergency resuscitation equipment and IM/IV epinephrine should be readily available. IM epinephrine should be used if there is hemodynamic instability or respiratory distress, but I am extremely sensitive to Epi, so use a low dose and push very slowly. Because of the risk of potentially lethal arrhythmias, IV epinephrine should only be administered in profoundly hypotensive patients or patients in cardio/respiratory arrest who have failed to respond to IV volume replacement and several IM doses of epinephrine. ***See attached anaphylaxis protocol*** Discuss serum mast cell tests in the event of a reaction.
- Protocol for hypoglycemic events and a plan to minimize length of NPO (nothing by mouth).
- Baseline low blood pressure (~85/55) and low blood volume. Care must be taken to maintain normal central venous pressure (CVP) and cardiac output and ensure anesthesia or hypovolemia don't cause dangerous hypotension. Bear in mind my normal low BP, so anaphylaxis is not suspected and treated when it isn't there. To guide induction and identify the depth of anesthesia, consider BIS monitoring. Consider an intra-arterial catheter for continuous blood pressure monitoring. When coming out of anesthesia, close monitoring is necessary to ensure volume shifts don't cause hypotension or tachycardia.
- Pain control: No opioids, narcotics or NSAIDS should be used.
- Possible airway difficulties:
    - Intubation should be done by fiberoptic or Glidescope because of craniocervical instability and decreased C-spine mobility.
    - Head and neck must be kept in a neutral position during surgery: hyperextension could cause spinal cord compression or subluxation with potential for neurologic injury.
    - Decreased oral opening, TMJ dysfunction.
    - Increased risk of bleeding from intubation and dysphagia from ETT (endotracheal tube) or LMA (laryngeal mask airway). Consider using smaller ETT/ scope (pediatric scope).

    - Keep airway pressures low due to increased risk of lung injury/pneumothorax or shunt with positive pressure ventilation.
- EDS surgery complications: Increased bleeding risk (if there is the possibility of a blood transfusion during elective surgery, consider autologous donation), fragile tissues that can tear easily, increased risk of perforation from scopes, increased risk for residual neuromuscular blockade, subluxations (also due to osteoporosis: head, neck, pelvis and limbs should be moved carefully and limbs kept close to body).
- Mast cell reactions to perfume, scents, cleaning products, tape, ECG electrodes, pain, stress, excitement, temperature extremes (important to keep patient warm), mold and *especially* hormones (menstrual cycle).
- Post-operative recovery accommodations, if needed:
    - Post-operative fluids.
    - Monitored for post-procedure anaphylaxis or mast cell reaction. Antihistamines and Prednisone should be used prophylactically to control reactions.
    - Discuss sensitivities to foods, temperature, sound, light, chemicals, fragrances, noise, sleep disruptions (request private room, quiet ward, door (versus curtain)). Bring your own safe food, soap, hand sanitizer, bedding etc.
    - Extra recovery time in hospital, if needed, or a reclining wheelchair/gurney during discharge from the hospital.