Hereditary angioedema (HAE) is a rare disease caused by a deficiency of C1 esterase inhibitor. It is characterized by recurrent episodes of swelling, without accompanying hives or purities, that often affects the skin or mucosa of the upper respiratory and gastrointestinal tracts, face and extremities. This swelling is often self-limiting, however the laryngeal edema associated with attacks may be life threatening. Various triggers have been reported including mild trauma, stress, medications or menstruation. In this case a 17-year-old female with a history of hereditary angioedema had frequent attacks characterized by laryngeal and abdominal edema. This case is significant because stress and pain were known triggers for her attacks.

Case Description
A 17-year-old 65 kg female with a history of hereditary angioedema, presented for surgery for a port placement secondary to her frequent episodes of angioedema. She reported having repeated episodes of airway swelling and abdominal swelling. She required the medication Cinrynze, a plasma derived C1 esterase inhibitor product, every three days. This medication is administered intravenously and given the patient had difficult IV access the decision was made to place a port. Her other medications included Firazyr, Ecallantide and an EpiPen on an as needed basis. She had to administer Ecallantide, a medication used for the treatment of HAE attacks given subcutaneously, which could be as frequent as once or twice a day. Prior to surgery she was premedicated with oral versed and lidocaine spray was used for IV start. She was then given Cinryze prophylactically one hour prior to surgery.

Discussion
Severe angioedema can occur in the perioperative period, causing fatal airway obstruction. It is important to understand and take the necessary precautions when managing patients with HAE. This case demonstrates that successful management of this rare condition can be obtained with administering prophylactic C1 esterase inhibitor and meticulous measures to avoid triggers.

References