EMERGENCY MEDICAL INSTRUCTIONS FOR PROFESSIONALS

WHEN DEALING WITH STIFF PERSON SYNDROME

Stiff person syndrome (SPS) is a rare neurological disorder characterized by the presence of fluctuating muscle rigidity and spasms of the trunk and more proximal body parts.

Patients with SPS may present as an emergency with severe pain and spasms of the lumbar paraspinal muscles and lower limbs. The spasms, often associated with intense pain, typically begin with an abrupt jerk followed by tonic activity that slowly subsides over seconds to, less commonly, minutes. Rarely, these spasms last days (status spasticus). The affected muscles are typically extremely hard to palpation with a board-like appearance, leading to hyperlordosis. Muscle spasms can occur spontaneously or be provoked by noise or movement. The spasms may be so forceful as to produce femoral fractures, joint subluxations and even herniation of abdominal contents.

Paroxysmal autonomic dysfunctions, such as transient hyperpyrexia, diaphoresis, tachypnea, tachycardia, pupillary dilation, and arterial hypertension, are recognized. Rhabdomyolysis can be a potential complication of the excessive muscle contractions in SPS. Sudden death can also occur due to an acute autonomic failure. In a significant number of cases, SPS is believed to be mediated by autoantibodies to glutamic acid decarboxylase (anti-GAD) that limit the gamma amino-butyric acid (GABA) neuronal activity and lower the threshold for muscle spasms, other neurologic and psychi-atric features of the disorder. SPS with elevated serum anti-GAD levels may occur with other autoimmune disorders, including diabetes mellitus, Graves' disease, Hashimoto's thyroiditis and pernicious anemia. Ten percent of cases with normal levels of this antibody may be related to au-toantibodies against amphiphysin which commonly rep-resent a paraneoplastic syndrome related to breast cancer, mediastinal tumors, small cell lung cancer, Hodgkin's disease and colon cancer.

The management of SPS is based on the use of drugs that promote GABAergic inhibition, for example, benzodiazepines (diazepam, alprazolam, and clonazepam) and baclofen. High-dose benzodiazepines can abolish the excessive motor unit activity. Oral baclofen provides relatively modest relief, while intrathecal administration seems to be much more effective. Finally, variable degrees of benefit have been reported with the use of antiepileptic drugs, such as valproic acid, levetiracetam and gabapentin.[1]

1. Munhoz RP, Moscovich M, Araujo PD, Teive HAG. *Movement Disorder Emergencies*. Arq Neuropsiquiatr. 2012 Jun;70(6):453-61. http://www.ncbi.nlm.nih.gov/pubmed/22699544
2. Baclofen Emergency Protocol http://www.maryfreebed.com/ITB
3. Baclofen Call Flowsheet http://www.maryfreebed.com/ITB
4. Benzodiazepine Toxicity http://emedicine.medscape.com/article/813255-treatment
5. Focus On: Best Practices for Seizure Management in the Emergency Department http://www.acep.org/Content.aspx?id=72861

Due to the rare nature of the disease, no specific technique has been recommended. The decision to employ either general anesthesia or regional block has to be made based on the type of surgery, involved anatomy, disease severity, patient preference and comfort level of the participating anesthesiologist.1

Total intravenous anesthesia has been reported to be affective. Regional anesthesia could be advantageous by providing effective and deep analgesia without necessitating the use of muscle relaxants and inhalation agents.

Challenges to spinal block include difficulty locating anatomical landmarks, limitations to proper positioning, and a chance of painful spasms and rigidity in response to the needle insertion. Presence of an intrathecal pump may necessitate a fluoroscopic guided neuraxial procedure.

It is crucial that the anesthesiologist be informed of the medications and dosages you are taking. The various forms of anesthesia drugs often fall in the GABAergic category. When combined with your daily medications, they can cause a fatal overdose or severe hypotonia.

There are no contraindications to any anesthetic agents or procedures. But due to the often high levels of muscle relaxers, patients should be monitored for hypotonia and the need for supportive ventilation.

Many anesthestic agents involve blockade of GABA receptors at various sites. Both inhalation agents and intravenous agents have the potential to cause hypotonia by causing GABA antagonism. They can increase the proportion of desensitized neuromuscular receptors and can result in a non-competitive blockade and prolonged duration of action. The depth of neuromuscular blockade should be closely monitored and the muscle paralysis must be adequately reversed, keeping in mind there may be no correlation between depth of muscle paralysis and the hypotonia observed in several case reports.

There is no effect direct effect of SPS or its therapy on the neuromuscular junction, therefor it should not affect the actions of nondepolarizing agents.

Patients may be on any combination of benzodiazepines, baclofen, gabapentin, vigabatrin, venlafaxin, plasmapheresis, IVIG, or high-dose corticosteroids.

It is important to cover with steroid prophylaxis to supplement for the possible cortisone suppression. Diazepam and baclofen therapy should be continued during the perioperative period. Discontinuation could cause severe withdrawal reaction. The anesthetist should be aware of and prepared to respond to such a reaction. The patient may need supplemental steroid therapy if they are on long-term steroid medication.

Skeletal muscle relaxants should be avoided as they can potentiate hypotonia. Use of muscle relaxants needs to be monitored and small doses of short acting relaxant should be titrated.

Though there is little research, case reports document resistance to succinyl choline.

If the patient has kyphosis, hyperlordosis, or contracture, care must be taken with positioning. The patient should be positioned with the appropriate aids or pillows.

The patients have a startle reaction to voluntary movement, emotional upset, or unexpected auditory, tactile, or somatic stimuli. Especially when prone, they should be well supported for rigidity and sudden spasms.

In addition to routine monitoring of blood pressure, oxygen saturation, ECG and endtidal CO2, the anesthetist should monitor for neuromuscular paralysis. There is potential for respiratory insufficiency due to muscular rigidity.

Bispectral index should be kept between 40 and 60, indicating the depth of anesthesia.

Although no association with malignant hyperthermia is suspected, it allows you to differentiate the possible complications including baclofen withdrawal and nerolept malignant syndrome.

1. Anesthesia recommendations for patients suffering from stiff-man synrome. Orphan anesthesia.net. http://www.orphananesthesia.eu/en/rare-diseases/published-guidelines/cat_view/61-rare-diseases/60-published-guidelines/98-stiff-person-syndrome.html