

## Abstract # 04

### The Use Of Dexmedetomidine As An Adjuvant To Regional Anesthesia In A Rare Case Of Adult Tay-Sachs Disease: A Case Report

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**Introduction:** Tay-sachs disease (TSD) is a rare inherited autosomal recessive metabolic disorder due to deficiency of beta-hexosaminidase A enzyme. The disorder leads to toxic accumulation of GM2-ganglioside in the nervous system causing destruction and death in children, and neurological, and intellectual impairment in adults. The so-called infantile onset form affects mostly Ashkenazi Jewish children, while the late onset (LOTS), or adult form designated as chronic GM2-gangliosidosis, does so to a lesser extent. Patients with LOTS develop neurological symptoms with advancing age including ataxia, unsteady gait, muscle weakness, and slurred speech. To date, no curative therapy for TSD exists, however research exploring gene therapy to reverse the CNS damage appears promising.

**Case Report:** A 55 y/o female with adult onset TSD, had a right knee replacement nine years prior. She developed neurological symptoms including gradually progressive bilateral upper and lower extremity, respiratory and bulbar muscle weakness. Family history was significant for amyotrophic lateral sclerosis with questionable malignant hyperthermia susceptibility for which the patient received genetic counseling. Other medical problems included COPD, neurogenic diabetes insipidus, temporal lobe epilepsy, hypothyroidism, and ADD. Past surgical history was significant for post-operative mechanical ventilation. She was scheduled for elective revision of knee replacement secondary to prosthetic joint implant failure. An epidural catheter technique was performed preoperatively using standard ASA monitors, supplemental oxygen via a nasal cannula. Regional anesthesia was induced with a combination of local anesthetics and opioids, while sedation was achieved using the alpha 2 adrenergic agonist dexmedetomidine as an adjuvant secondary to respiratory muscle weakness, using a continuous infusion of 1.5-2 µg/kg/h, supplemented with Midazolam .1mg/kg. The level of sedation was guided by the BIS monitor at 65-75. Hemodynamics remained stable during the operative course with a transient hypotensive episode requiring vasopressor support secondary to local anesthetic administration. The patient was transferred to the post-anesthesia care unit where recovery was uneventful. She had excellent post-operative analgesia via the epidural catheter, enabling her to undergo successful rehabilitation.

**Conclusion:** There are no cases to date describing the anesthetic management of a patient with adult onset TSD. This case report demonstrates the safety and efficacy of using dexmedetomidine as an adjuvant to a regional anesthetic technique to provide sedation, analgesia and maintain airway reflexes. More research is warranted to determine the safety of the use of dexmedetomidine and other regional anesthetic techniques in such disorders.

**References:** D. Soliman, M.D., A. Maslow, M.D. Dexmedetomidine: A Review of Its Benefits and Applications. SCA Newsletter, April 2006, Vol. 5, No. 2, P. 8-9.