

An Interdisciplinary Team Approach to Sedations for Spinraza Intrathecal Injections in Patients with Spinal Muscular Atrophy

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Introduction: Spinal Muscular Atrophy (SMA) is a rare autosomal recessive neuromuscular disorder. SMA presents in the form of progressive muscle wasting, mobility impairment, and has the potential to cause respiratory compromise. Spinraza is an antisense oligonucleotide that has been FDA approved for intrathecal administration since December 2016, and is the only treatment available for this traumatic disease. Spinraza is changing the prognosis of patients with SMA and improving their quality of life. Four initial loading doses are administered, then children graduate to therapy every four months for the duration of their lifetime. We've outlined the methods of the interdisciplinary team for sedating patients with SMA.

Discussion: The Pediatric Pain Sedation and Consult service (PSC) operates in a 150-bed children's hospital within a large academic center. The PSC service consists of specially trained Registered Nurses, Child Life Specialists, Pediatric Nurse Practitioners, and Pediatric Anesthesiologists. Our service provides general anesthesia, moderate sedation, or anxiolysis for these patients to undergo lumbar punctures for Spinraza to be administered. The use of anesthesia during this procedure helps to reduce anxiety and pain, as well as increase the success rate of lumbar puncture in this population. All patients are triaged each visit to assess their progression of SMA and respiratory status. This assessment determines which form of anesthesia will be the most safe and effective for each patient. For the first lumbar puncture, patients are assessed and receive no sedation, moderate sedation or general anesthesia by the Pediatric Anesthesiologist to ensure patient safety. At subsequent visits, if the patient requires only moderate sedation or anxiolysis, they may be cared for by the Pediatric Nurse Practitioner and Registered Nurses. Many patients, who are not respiratory compromised, are able to complete their lumbar puncture with moderate sedation or anxiolysis.

References:

1. Hache, M et al., Journal of Child Neurology, 2016