A Comprehensive Institutional Overview of Intrathecal Nusinersen Injections for Spinal Muscular Atrophy

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Introduction

Spinal muscular atrophy (SMA) is an autosomal recessive neuromuscular disorder resulting in progressive muscle weakness. In December 2016, the U.S. Food and Drug Administration approved the first ever treatment for SMA, a drug named nusinersen (Spinraza) which is administered intrathecally. However, many SMA patients have neuromuscular scoliosis and/or spinal instrumentation resulting in challenging intrathecal access. Therefore alternative routes must be considered in these complex patients.

Research Question

Can a decision tree be created for physicians to utilize when administering nusinersen (Spinraza) to children with Spinal Muscular Atrophy, even with complex spinal anatomies and hardware?

Materials and Methods

This study was reviewed and approved by our institution's institutional 13 review board. From March to December 2017, institutional SMA patients were referred for intrathecal nusinersen injections. In select patients with spinal hardware, spinal imaging was requested to facilitate pre-procedure planning. Standard equipment for intrathecal injections was utilized. All patients were followed-up by their referring neurologist.

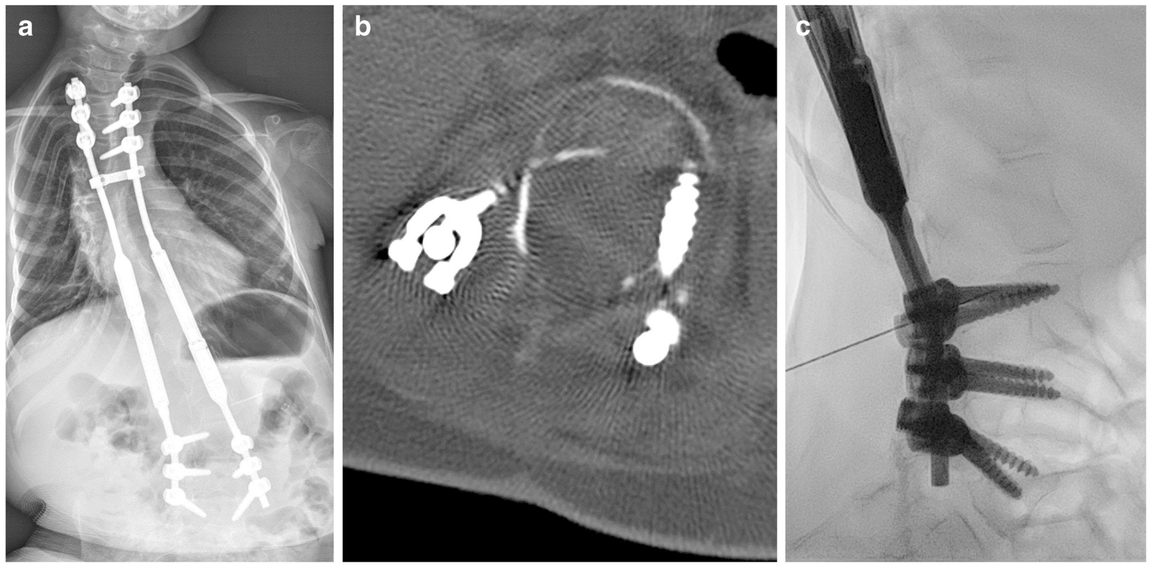

Figure 1: Spinal muscular atrophy type 2 and spinal hardware in a 7-year-old girl. a Anteroposterior radiograph demonstrates scoliosis post repair and neuromuscular disease. b Axial image from pre-procedural non-contrast diagnostic lumbar spine CT demonstrates patency of the posterior spinal canal, with lack of bony fusion, at the level of the most cephalad transpedicular screws. c Lateral fluoroscopic image demonstrates spinal needle tip superimposed over the most cephalad transpedicular screws, positioned in the subarachnoid space at L2-L3 via an interspinous approach.

Results

A total of 104 intrathecal nusinersen injections were performed in 26 patients with 100% technical success. 60 procedures were performed without pre-procedural imaging and via standard interspinous technique. The remaining 44 procedures were performed in 11 complex (i.e. neuromuscular scoliosis and/or spinal instrumentation) patients requiring pre-procedural imaging for planning purposes. 19 of 44 complex procedures were performed via standard interspinous technique from L2 to S1. 22 of 44 complex procedures were performed using a neural-foraminal approach from L3 - L5. 3 of 44 complex procedures were performed via cervical puncture technique. There were no immediate or long-term complications but 1 short term complication of meningismus and back pain at the injection site.

Conclusion

Our early experience with nusinersen administration has led to the development of an algorithm (Fig. 2) to assist in promoting safe and effective nusinersen administration in children with spinal muscular atrophy regardless of SMA type, abnormal spinal anatomies and complex spinal instrumentation. Our 100% technical success of intra-thecal access suggests that more aggressive surgical approaches, such as reservoir placement or creation of a surgical window via a bone graft, should be reserved for children with no safe percutaneous access routes.

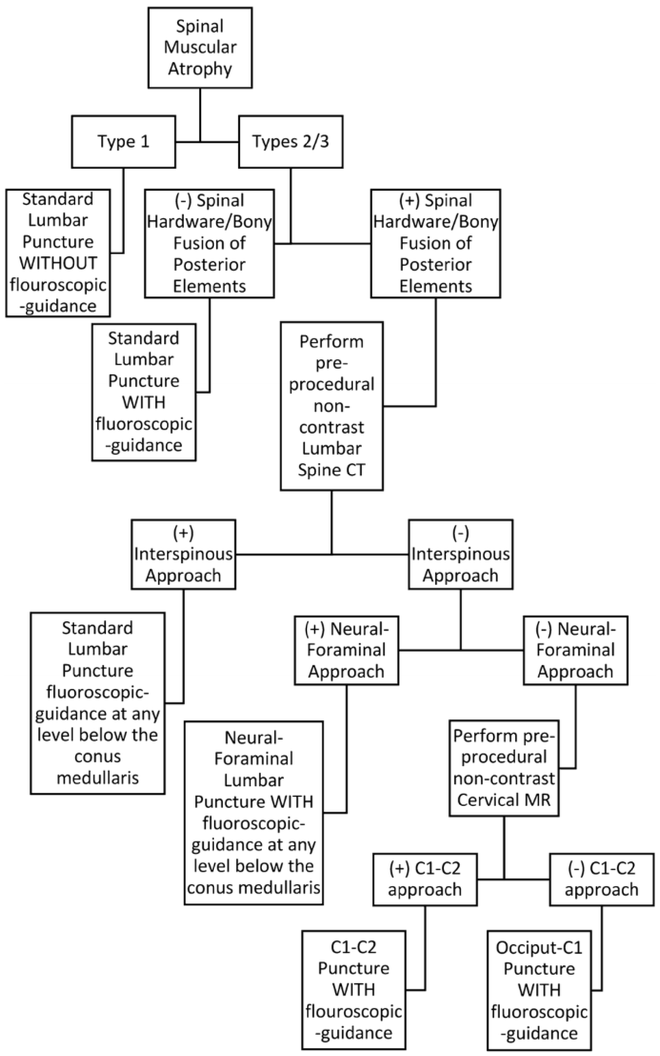

Figure 2: Approach to intra-thecal therapy in children with spinal muscular atrophy (SMA). The approach to the child with SMA depends on the status of the bony spine. Children without scoliosis, spinal fusion or instrumentation (i.e. simple spine) are candidates for non-image-guided standard interspinous lumbar puncture. In contrast, children with a complex spine require pre-procedural imaging evaluation for route planning and in most cases intra-procedural image guidance

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