Spinal Muscular Atrophy

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Abstract: Spinal muscular atrophy (SMA) is a progressive neuromuscular condition characterized by hypotonia. Recent advances in the medical treatment of SMA have increased life expectancy and improved functional abilities. The myriad of exciting new medical treatments will complicate the study of orthopaedic pathology in these patients, and the "natural history" will be continually changing. As a result, orthopaedic management of scoliosis, hip subluxation/dislocation, joint contractures, and insufficiency fractures in SMA is likely to take on a larger role. Osteopenia is reported to be the most severe in SMA above all other neuromuscular conditions. Some patients with SMA have unique parasol chest deformity that contributes to the challenge of managing spine deformity. In addition, intrathecal administration of disease-modifying agents requires access to this anatomic space and must also be considered during posterior spine surgery. Emerging evidence suggests that hip dislocation is painful in some SMA patients, and a better understanding of who is at risk for hip pain and how best to manage these patients is needed. In light of the recently evolving expectations for life expectancy and functional abilities, this review offers an overview of the recent evidence in the orthopaedic management of SMA.

Key Points:
- Recent development of disease-modifying agents, including nusinersen (Spinraza®) and onasemnogene abeparvovec-xioi (Zolgensma®), increase life expectation, and improve functional ability in patients with spinal muscular atrophy (SMA).
- Treatment of orthopaedic manifestations of SMA, including scoliosis, hip subluxation/dislocation, joint contractures, and insufficiency fractures, is likely taking on a larger presence with improved medical management and development of disease-modifying agents.
- Management of spinal deformity in SMA must consider severe osteopenia, altered vertebral anatomy (short pedicles and elongated vertebral bodies), and parasol chest deformities associated with rib overhang.
- Hip dislocation is likely painful in a subset of patients with SMA and is an indication for hip reconstruction.
- Joint contractures and insufficiency fractures share a common etiology of muscle pathology (stiff and nonfunctioning) and may be addressed with surgical lengthening, increased weight-bearing, and bisphosphonates.

Introduction

Spinal muscular atrophy (SMA) is a common, fatal autosomal recessive condition, occurring with a reported incidence between 1/6,000 to 1/10,000 in a European population.[1] It is a progressive neuromuscular disorder affecting the anterior horn cells of the spinal cord and presents with weakness and delay or regression of motor milestones. The diagnosis of SMA should be considered in children presenting with hypotonia, absent deep tendon reflexes, muscle weakness in both legs and arms, and fasciculations. These findings should prompt a referral to pediatric neurology as well as an *SMN1* gene

deletion test, which is typically a blood test.[2] Prenatal testing is also now available.[3] With the recent disease-modifying drugs, many states are moving towards SMA testing in the newborn screening panel.[4] Creatine kinase, nerve conduction studies, and muscle biopsy may also be considered to rule out other degenerative muscle disorders. Once the diagnosis is made, a multidisciplinary team of specialists should be involved in the care of the patient, including a pediatric neurologist, pulmonologist, gastroenterologist, and physical and occupational therapy.

SMA type	Age of Onset	Motor Milestones	Average Age of Death	Eponym
1	< 6 months	Unable to sit without support	< 2 years	Werdnig-Hoffman
2	<18 months	Sit independently, cannot stand	2nd through 3rd decade	Dubowitz
3	> 18 months	Stand and walk independently	Normal life expectancy	Kugelberg-Welander

Table 1. Classic types of spinal muscular atrophy

Patients with SMA have a wide spectrum of phenotypes ranging from death during early childhood due to respiratory failure to normal life expectancy with ambulatory function. The traditional classification for SMA differentiates patients based on the age of presentation and offers a prognosis for life expectancy and motor milestones[2,5] (Table 1). Type 1 SMA (Werdnig-Hoffman disease) manifests prior to six months of age. Patients are unable to sit, and without aggressive multimodal medical management, they have a life expectancy of less than two years. In Type 2 SMA (Dubowitz syndrome), symptoms start prior to 18 months of age. Patients are able to sit but not stand independently. Patients typically live into the second or third decade of life. Type 3 SMA (Kugelberg-Welander syndrome) is the mildest form with age of onset after 18 months of age. Patients stand and walk independently and have a normal life expectancy.

New disease-modifying agents, including nusinersen (Spinraza®) and onasemnogene abeparvovec-xioi (Zolgensma®), offer significant promise for changing the natural history of SMA.[6] These medications, in combination with aggressive multidisciplinary medical management, increase overall survival.[7,8] With increased survival, the orthopaedic surgeon plays a larger role in the care of patients with SMA. Common orthopaedic manifestations include spinal deformity, hip subluxation/dislocation, joint contractures, and insufficiency fractures. Areas of recent controversy in the orthopaedic literature include the impact of spine surgery on pulmonary function as well as the role of hip reconstruction in dislocated hips. This review focuses on the current paradigm for orthopaedic management in SMA.

Etiology

SMA is transmitted in an autosomal recessive manner and is caused by a homozygous deletion of exon 7 and/or exon 8 in the survival motor neuron 1 (*SMN1*) gene, located on chromosome 5q13.[9,10] The survival motor neuron protein is highly expressed in the spinal cord and brainstem and is important in the development of dendrites and axons.[11] In SMA, inadequate expression of survival motor neuron protein results in the degeneration of anterior horn cells of the spinal cord. This pathology in the anterior horn cells causes classic lower motor neuron findings, including global flaccid paralysis as well as fasciculations.

SMN2 is a paralogous gene also located on chromosome 5q and is present in variable copy numbers.[12] *SMN2* produces a splice site variant which results in exclusion of exon 7 and ultimately a truncated and dysfunctional

survival motor neuron protein;[13] however, due to inherent splicing errors, *SMN2* can produce full-length active survival motor neuron protein, only at levels 5-10% of that produced by *SMN1*. Thus, an increased number of copies of the *SMN2* gene may lead to increased levels of survival motor neuron protein. However, a direct relationship between the number of copies of *SMN2* gene and SMA phenotype does not necessarily exist.[14,15] Many of the new disease-modifying agents target these genetic pathways.

Disease-Modifying Agents

Over the past twenty years, aggressive multi-disciplinary treatment of children with SMA demonstrated that the natural history and traditional life expectancies could be dramatically altered.[7] More recently, there has been growing interest in disease-modifying agents for SMA developed both for the promise of effective treatment provided to families as well as the cost associated with these drugs. Nusinersen (Spinraza®) was approved by the U.S. Food and Drug Administration (FDA) in 2016 for the treatment of SMA.[16] Delivered intrathecally, nusinersen is an antisense oligonucleotide that binds to a specific sequence within the SMN2 pre-messenger RNA. Binding of nusinersen in this region modifies the splicing of the SMN2 pre-messenger RNA, which results in increased expression of full-length survival motor neuron protein.[17] Therapy with nusinersen demonstrated improved survival and motor development in patients with severe SMA treated between 30 and 262 days of age and between two and nine years of age.[18]

Onasemnogene abeparvovec-xioi (Zolgensma®) is a gene therapy medication approved by the FDA in May 2019 for the treatment of any type of SMA.[19] The medication is a viral vector consisting of the AAV9 viral capsid, which contains the SMN1 transgene, delivered as
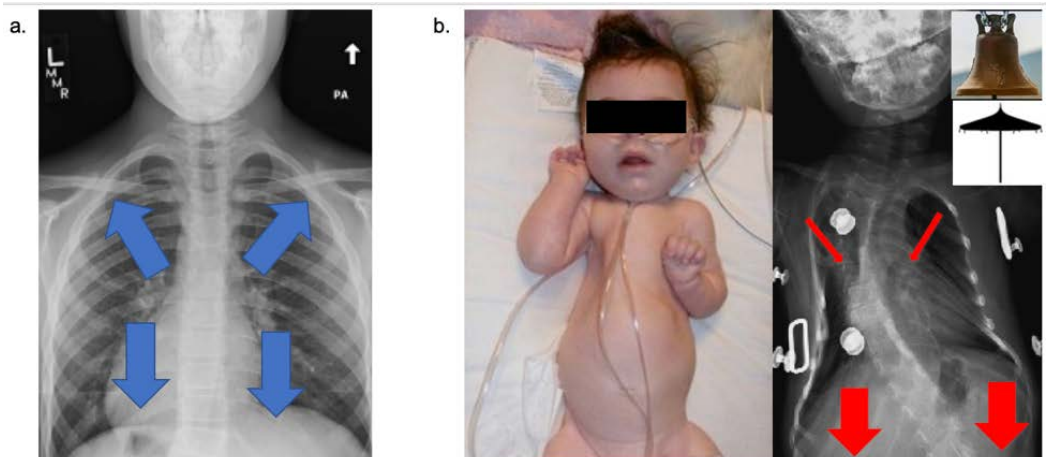

Figure 1. Chest deformity in spinal muscular atrophy.
a. Under physiologic conditions, the intercostal muscles balance the pull of the diaphragm, preserving the size and shape of the thoracic cavity. b. In spinal muscular atrophy, weak intercostal muscles and relative overpull of the diaphragm results in rib collapse and the characteristic bell-shaped or parasol (umbrella) deformity.

a single intravenous dose. The viral vector delivers the SMN1 transgene, which results in increased production of survival motor neuron protein. In a single-arm trial consisting of 12 patients with type I SMA receiving high dose Zolgensma®, all patients were alive at two years, compared to an 8% survival rate in a historic cohort.[20] In addition, eleven patients were sitting independently, nine rolled independently, and two were walking at the 24-month post-treatment visit. Seven of ten patients who did not require baseline respiratory supportive care remained free of respiratory supportive care. These remarkable results were followed by FDA approval of Zolgensma® for children under the age of two with SMA.

Controversy and popular media attention over these medications for an otherwise rare disease stem from the cost. Zolgensma® costs U.S. $2.125 million per dose, making it the most expensive medication in the world as of 2019.[21] Nusinersen costs U.S. $125,000 per dose.[22] Five to six doses are required in the first year of treatment (U.S. $625,000 to $750,000) followed by three doses annually after the first year (U.S. $375,000).
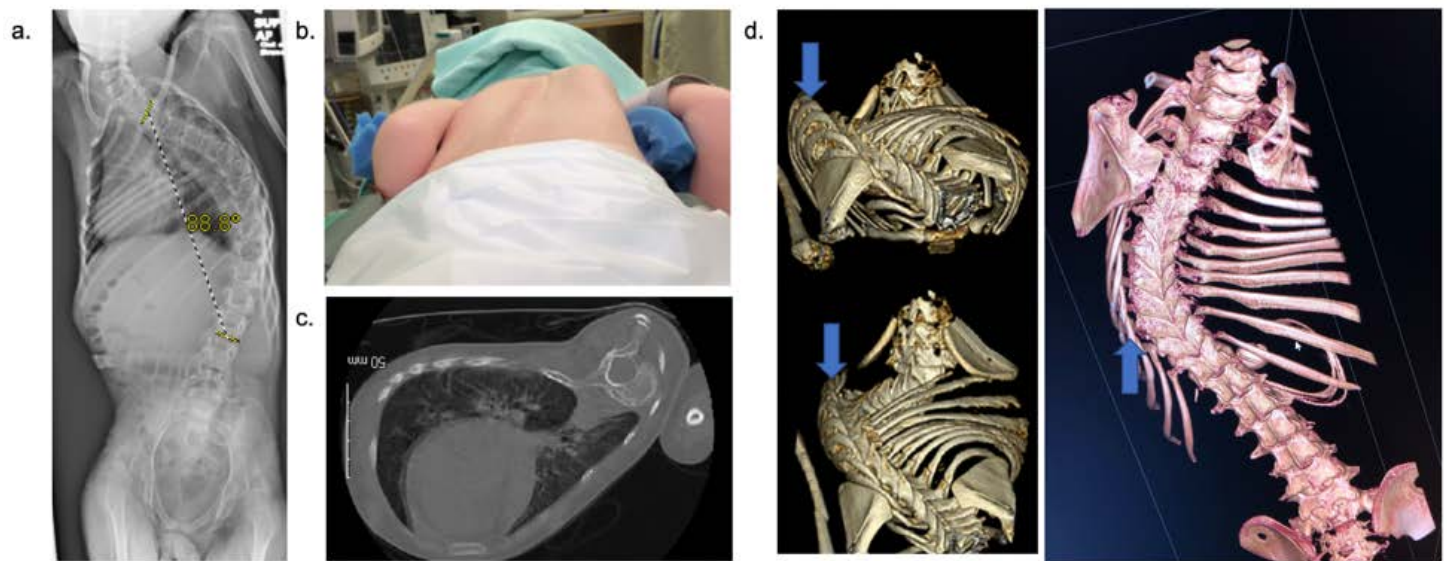
Figure 2. Spine and chest deformity in spinal muscular atrophy. *a. Radiograph of a patient with 88° long, sweeping curve characteristic of spinal muscular atrophy. b. Intraoperative photograph of patient in prone position demonstrates the significant "knife-edge" rib deformity on the convex side of the curve. c. Axial CT scan represents the significant vertebral rotation and severe rib deformity. d. Three-dimensional CT reconstruction highlights the rib shingling on the convexity of the curve (blue arrows).*[27]

Efforts were made by the manufacturer of Zolgensma® to provide the medication for free to 100 patients across the world determined by a lottery-style program, but this action was met with significant controversy due to questions of the ethics of selecting patients in a lottery.[21] In addition, data manipulation by the pharmaceutical companies producing Zolgensma® contributed to public uproar.[23]

Management of Spinal Deformity

Prior to the introduction of disease-modifying drugs, nearly all children with Type 1 and Type 2 SMA developed scoliosis[24] and was reported to be one of the most concerning aspects of SMA among healthcare professionals and families of patients with SMA.[25] Curves are often long and sweeping, similar to scoliosis associated with other neuromuscular conditions. However, development of the classic parasol or bell-shaped chest deformity[26] (Figure 1) is a unique feature in the SMA population that can be seen with or without the spinal deformity.

This deformity is thought to develop due to weak intercostal muscles and a relatively strong diaphragm. These abnormal forces pull the ribs down, causing the ribs to sag and collapse. When combined with scoliosis, the sagging ribs create a characteristic thoracic appearance. On the convex side of the curve, the ribs shingle lying on one another and abut the vertebral bodies. The stacking of the ribs in this manner creates a narrow bony "knife's-edge" ridge immediately adjacent to the spine (Figure 2).

Scoliosis treatment options are generally limited to growth-friendly or definitive fusion techniques. Derotational casting in early onset scoliosis (EOS) SMA patients is not typically recommended due to associated pulmonary morbidity. Bracing is also challenging in this patient population due to weakness of respiratory muscles and associated restrictive lung disease. The literature reports mixed success for slowing the progression of scoliosis in this patient population.[27] The senior author recommends semi-rigid bracing in patients with a curve greater than 20° who can tolerate being upright either in a wheelchair or stander. The orthosis is
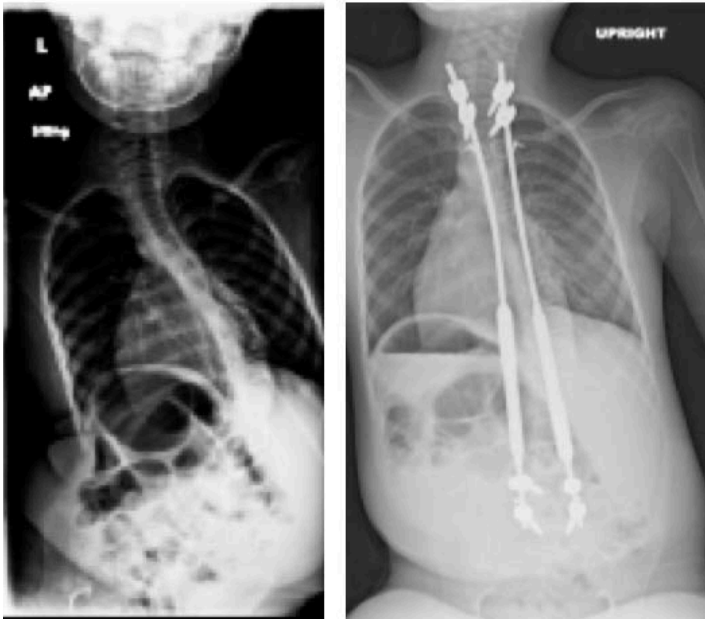
Figure 3. *Magnetically controlled growing rods in early onset scoliosis in spinal muscular atrophy. Growth-friendly construct achieved adequate correction with instrumentation from the upper thoracic spine to L5 and facilitated growth.*

more for the postural support, than to prevent curve progression. Brace wear is more sporadic in patients with Type 1 SMA. Braces are generally semi-rigid thoracolumbar sacral orthoses with a large abdominal cut-out to facilitate diaphragmatic breathing and allow access to gastrostomy tubes (G-tubes).

Progressive deformities with Cobb angles greater than 50-60° is an indication for surgery for patients in all age groups.[29,30] Allowing the deformity to worsen past this magnitude adds technical challenges to the surgery and is thought to increase the risk for complications. Although strict guidelines for the timing of definitive fusion do not exist in the literature, as a rule of thumb, children younger than eight years of age undergo growth-friendly techniques. Children older than ten years of age undergo definitive posterior spine fusion. In patients between these age groups, patient-specific factors must be considered with either definitive fusion or growth-friendly techniques available as options.

Aggressive medical treatment and disease-modifying agents have increased survival in Type I SMA patients, and most will develop early onset scoliosis (EOS). Growth-friendly techniques, including vertically expandable prosthetic titanium ribs, Luque Trolley techniques, and traditional growing rods, have shown good results.[31–33] Magnetically controlled growing rods also show significant promise in the management of EOS in patients with SMA (Figure 3).

Another potential benefit of traditional posterior growing rods is that conversion of these patients after lengthening to definitive posterior fusion does not appear to be necessary. In a retrospective review of 12 patients with SMA and EOS who underwent growth friendly techniques, only one patient underwent conversion to definitive fusion.[34] Definitive fusion was performed in this patient to improve hip pain; however, hip pain persisted postoperatively. Another patient had occult failure of a rod that did not require revision. The authors concluded that these patients may not automatically require definitive posterior fusion and that such a decision could be made on a case-by-case basis.

Similar to patients with other neuromuscular conditions, posterior spinal fusion appears to benefit adolescents with scoliosis. Seating balance is improved, and

Figure 4. ***Altered osseous anatomy of vertebral body in spinal muscular atrophy***. *Lateral radiograph demonstrates elongated vertebral body, which must be taken into account when selecting pedicle screw length.*[27]

correction of curve magnitude is maintained. The impact on pulmonary function is equivocal.[32,34-37] In a systematic review, Howard et al. found that surgery did not reliably improve pulmonary function compared to baseline.[37] However, they rated this recommendation as Grade C based on Level IV and V evidence. Spinal fusion has, however, been associated with functional loss as some patients lost either ambulatory or sitting function.[38] Nevertheless, overall, spine surgery was found to improve parental quality of life and family impact.[39]

Often, patients with SMA undergoing surgery for spinal deformity are medically fragile and are thought to be at high risk due to their pulmonary status. We have found that a robust multi-disciplinary approach to medical perioperative management (Table 2) allows for these procedures to be safely performed. Our protocol typically institutes pre/intra-operative total parental nutrition (TPN) in patients with Type 1 SMA and in select, weaker patients with Type 2 SMA. Anesthesia should be made aware that roughly one quarter of these patients will have a difficult intubation. Given the frailty of these children and their often-smaller size, additional anesthetic time at the beginning and end of each of these cases should be anticipated. Postoperatively, we recover all of our children in the ICU, and our average length of stay is 7-8 days regardless of SMA type. Atelectasis and ileus are the most common postoperative complications. Using this protocol, pneumonia, and the need for reintubation are rare in our experience. We conclude that surgical treatment of spinal deformity in SMA can be performed safely when appropriate perioperative measures are taken.

Several technical considerations should be recognized in these challenging cases. First, patients frequently have upper and lower extremity contractures and osteopenia. Care must be taken during positioning to avoid fractures. Intra-operatively, this osteopenia can contribute to screw plow and pullout during deformity correction. This may be prevented by utilizing less rigid constructs, including polyaxial screws, supplemental sublaminar wiring, and
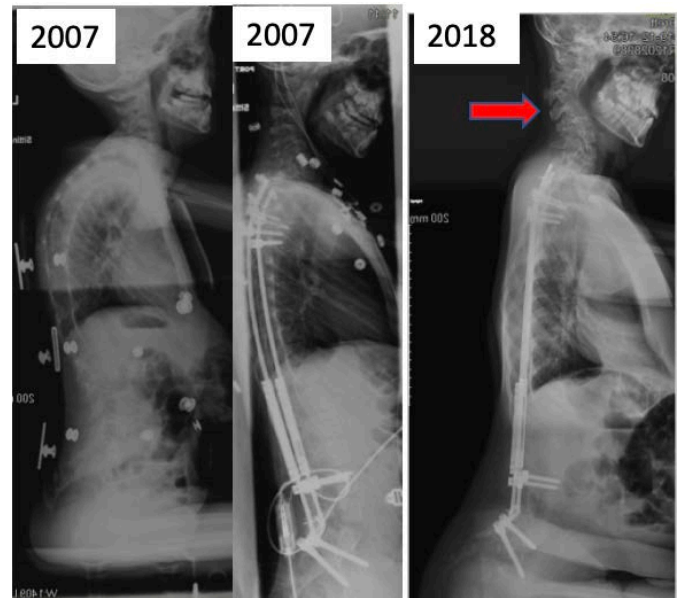

Figure 5. Cervical kyphosis after spine surgery.
Preoperative and postoperative lateral radiographs of the spine demonstrate decreased thoracic kyphosis after surgery with a progressive increase in cervical kyphosis 11 years after spine surgery (red arrow).
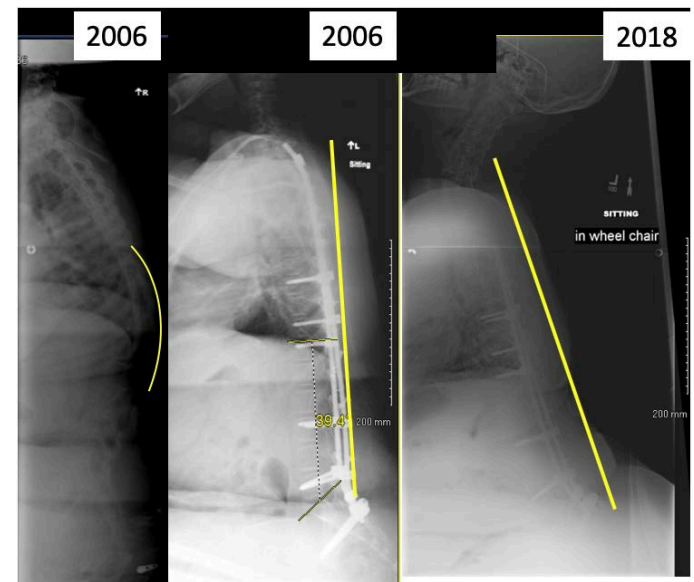

Figure 6. "Tipped trunk" appearance after spine surgery.
Preoperative lateral radiograph of the spine demonstrates thoracic kyphosis (curved yellow line). Decreased thoracic kyphosis is present on immediate postoperative radiograph in 2006 (yellow line). At 12-year follow-up, the patient has a "tipped trunk" appearance (yellow line) with positive sagittal balance and parallelism between the sacrum and the femur.

less stiff rods. Second, hip pain may be associated with pelvic fixation in those patients with dislocated hips. As such, fixation is usually stopped short of the pelvis at L5 as long as a balanced pelvis can be achieved. This practice is supported by case series showing adequate control of pelvic obliquity after stopping at L5.[40,41] Both pedicle screw and sublaminar wire fixation may be required at L5 if instrumentation stops at this level to ensure adequate fixation. Alternative fixation options should also be available at other levels, including hooks and sublaminar wires. Frequently, these patients have tall, narrow vertebral bodies, which accommodate short pedicle screws, sometimes less than 20mm in length (Figure 4). A preoperative CT scan defines pedicular anatomy and helps ensure that appropriate length pedicle screws are available.

Finally, it is important to provide a means for easy intrathecal access that can be used for later drug delivery. Various techniques are available, including laminotomy,[42] cannulated screw placement, and creating bone voids in the midline of a fusion at several locations by intentionally avoiding bone grafting in these areas or placing fat grafts or Gelfoam. Others have sought to place novel subcutaneous catheters consisting of an implantable infusion port connected to a baclofen pump catheter tunneled subcutaneously into the intrathecal space.[43]

SMA patients have postoperative risks similar to those observed in other neuromuscular conditions. Patients are at risk of pulmonary and wound healing complications.[45-46]

While mechanical complications after treatment of scoliosis is relatively uncommon due to the lower functional demands in this patient population, recent observations reveal that secondary sagittal plane deformities (including cervical kyphosis and trunk
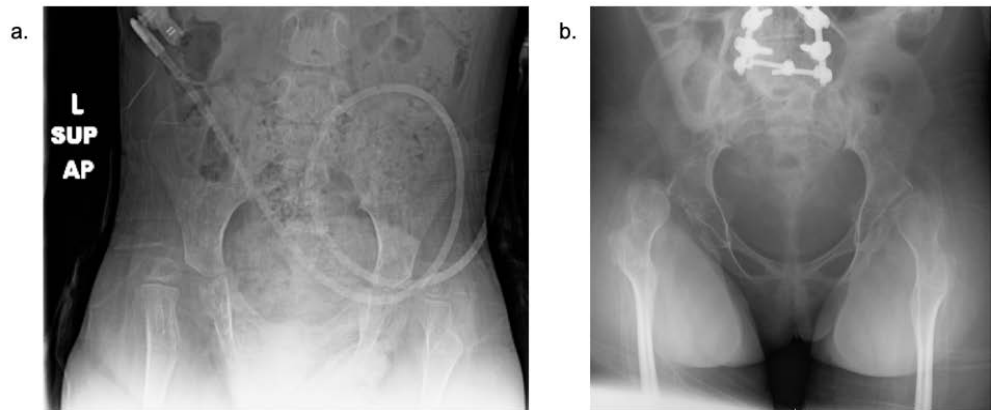

Figure 7. Hip pathology in spinal muscular atrophy (SMA). a. Bilateral hip dislocation in a patient with Type 1 SMA. b. Unilateral right hip dislocation and left hip subluxation with acetabular dysplasia and proximal femoral valgus in a patient with Type 2 SMA.

imbalance) may be common in these children following spine surgery (Figure 5).

Cervical myelopathy may be associated with kyphosis, requiring treatment with anterior cervical discectomy and fusion.[45] Furthermore, patients may develop excessive lordosis in the lumbar spine, leading to a "tipped trunk" in which the sacrum is parallel to the femurs (Figure 6).

Hip Management

Hypotonia associated with SMA alters normal muscular balance around the hip, leading to hip subluxation and dislocation[46] (Figure 7). Hip subluxation or dislocation is common. In one study, 62% (30/48) of hips were dislocated or subluxated in Type 2 SMA, and 29% (7/24) of hips were dislocated or subluxated in Type 3 SMA.[47] Patients maintaining ambulatory function are more likely to have a concentric hip than nonambulatory patients.[48] Hip dislocation is more likely to occur on the side with elevation of the pelvis.[49]

While it is generally accepted that hip reconstruction is indicated in patients who maintain ambulatory function and have ad equate muscle tone and function, previous orthopaedic literature suggested that hip dislocations in
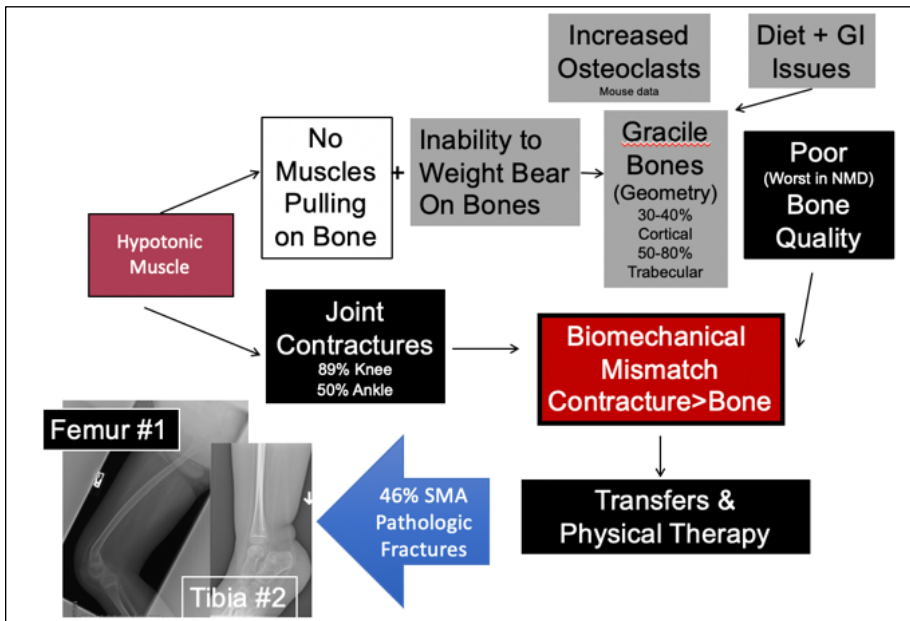
Figure 8. Pathophysiology of joint contractures and insufficiency fractures. *Hypotonic muscle is the common link for joint contractures and insufficiency fractures. Hypotonic muscles do not produce sufficient pull on bone, and patients frequently cannot bear weight. As a result, patients develop gracile bones, which is exacerbated by increased osteoclast activity as well as poor diet and gastrointestinal issues. Furthermore, hypotonic muscles result in joint contractures. The combination of joint contractures and bone osteopenia can lead to fractures during everyday activities, such as transfers and physical therapy.*

Predicting which children will have pain is difficult and ongoing work is being done to try and identify risk factors.

Most published studies have recommended against surgical intervention in nonambulatory children with SMA. However, the increasing life spans may uncover an increased incidence of hip pain that develops later in this population. Similarly, the increased muscle strength and tone combined with the increased life spans offered by the new disease-modifying treatments, may either decrease the incidence of hip dislocations or might convert the relatively painless hypotonic hip dislocations into more typical painful hip dislocations seen in patients with other neuromuscular dislocations. As such, the group from Boston Children's, including the pioneering work of Dr. Brian Snyder, have suggested improved long-term outcomes performing surgical reconstructions and have minimized previously reported complications by instituting a protocol of preoperative bisphosphonates, performing combined pelvic and femoral osteotomies, and using newer locking plate fixation. As medical treatment continues to advance, hip reconstruction in these patients may be performed more frequently.

Joint Contractures and Insufficiency Fractures

Joint contractures and insufficiency fractures in SMA are common and have related pathophysiologic mechanisms (Figure 8). Anterior horn cell pathology and hypotonia result in stiff muscles and joint contractures; 89% of patients with SMA have knee contractures, and 50% of patients with SMA have ankle contractures.

Bone mineral density in children with SMA was reported to be the lowest among neuromuscular disorders.[51] The absence of muscular activity in SMA, as well as the inability to bear weight, results in abnormally

patients with SMA were painless.[47] In addition, several reports conclude that reduction of hip dislocations is unlikely to maintain concentricity postoperatively.[46–49] Other authors indicate that the negative attributes of dislocated hips in spastic neuromuscular conditions, such as pain, difficulty with perineal care, and sitting imbalance, may not be present in conditions with low tone such as SMA.[50]

Currently, the treatment of hip pathology in the orthopaedic literature remains conflicted in the nonambulatory child. In our experience, the vast majority of nonambulatory children do not experience significant hip pain; however, a small subset of children with a Type 2 phenotype, do develop significant pain.

weak bones. These factors are further exacerbated by increased osteoclastic activity demonstrated in animal studies of SMA.[52] Finally, altered GI motility can further contribute nutritionally to the weakened bones. Pathologic fractures occur when these thin and weak bones are combined with joint contractures (Figure 9). Tibial and femoral fractures are the most common, with approximately half of children with SMA experiencing a pathologic fracture at some point in their life.

Historically, treatment for both contractures and insufficiency fractures was reactive. Joint contractures were addressed with physical therapy,[53] and fractures were treated with splinting for comfort.[54] Current treatment approaches are becoming more prophylactic in nature. Bone strength has been improved with bisphosphonate therapy. In a retrospective review of the impact of bisphosphonates on bone quality in patients with SMA (75% Type I), Nasomyant et al. found a trend toward improved bone mineral density at one-year follow-up and less fractures (1.4 to 0.1 fractures per year) in patients with more than two-year follow-up.[55] Patients received either pamidronate or zoledronate infusions starting at a median age of 6.7 years, with an average of approximately five infusions per patient. Furthermore, surgical intervention may be implemented for lengthening of joint contractures to facilitate weight-bearing, which improves bone quality.[56]

Summary

To conclude, this review offers a current perspective on the orthopaedic management of SMA with updates on the management of spine deformity, treatment of hip dislocation, and the use of bisphosphonate therapy. As patients have an increased lifespan with aggressive medical management and development of disease-modifying agents, the ability to evaluate outcomes after orthopaedic interventions will increase. However, with the advent of genetic testing and medical interventions, stratifying patients using the classic definitions of Types
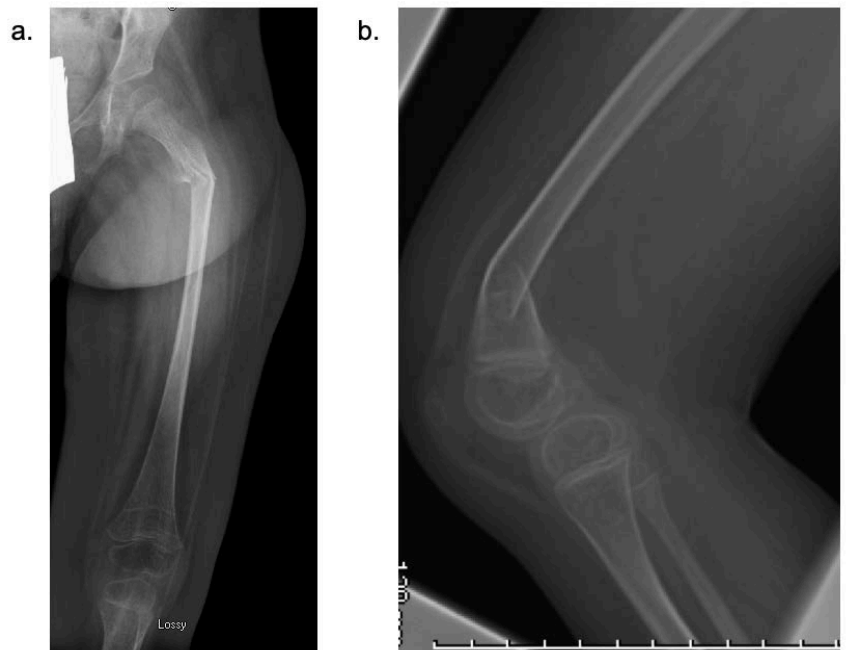
Figure 9. Pathologic fractures in spinal muscular atrophy. a. Proximal femoral fracture in a patient with spinal muscular atrophy. b. Distal femoral fracture in a patient with spinal muscular atrophy. Note the long, gracile architecture of the femur.

1, 2, and 3, including the age of onset, becomes more difficult. As patients may undergo different individual or combined therapeutic treatment at different ages, they may have differing responses. Thus, each of these medical advances may prove to be significant future confounding factors in trying to study orthopaedic outcomes. The ever-growing number of new therapeutics will have the effect to create more heterogeneity in this already small population, making the number of subjects that are truly "alike" even smaller. As a result, statistical comparisons of the outcomes of orthopaedic interventions will become even more challenging, especially for single centers. Perhaps in the future, this heterogeneity may be resolved by utilizing the highest or current functional status of patients, similar to the Gross Motor Functional Classification System in cerebral palsy.[57] The functional status may offer more information about prognosis and decision making for treatment rather than classic SMA types, copies of the SMN genes, or medical treatment history.

Table 2. *Perioperative Protocol for Spine Surgery in Spinal Muscular Atrophy*[27]

	Pre-operative consultation	Intraoperative	Post-operative
Pulmonary	• Pulmonary consultation for all patients prior to surgery • Conduct pulmonary function testing if patient is able to perform • Consider preoperative training on non-invasive positive pressure ventilation (NIPPV) if not currently managed with respiratory support during sleep due to increased risk for hypoventilation post operatively secondary to pain management and anesthesia • Consider preoperative training with mechanical insufflation-exsufflation if history of ineffective cough, recurrent pneumonia or low MEP (i.e., MEP < 60 cm H2O)	• Respiratory support per endotracheal intubation with positive pressure ventilation • Neuromuscular patients with chronic respiratory insufficiency require ventilatory support and not oxygenation alone, during induction and recovery. Patients should be kept intubated post operatively or extubated to NIPPV • Monitoring during anesthesia should include capnography to complement oximetry to avoid hypercarbia and hypoxemia.[3]	• Implement airway clearance post operatively with secretion mobilization technique preferably intrapulmonary percussive ventilation (IPV, Metaneb) followed by mechanical insufflation-exsufflation and airway suctioning within 1 hour of admission to post operative care unit and every 4 hours post operatively via ETT, tracheostomy tube or orally. • Consider delaying extubation until respiratory secretions well controlled, weaned to room air and pain control is optimized. • Optimize ventilation before adding oxygen as the most likely cause of hypoxemia is hypoventilation. Hypoventilation is exacerbated by narcotic pain use. • Extubate patients to NIPPV and work toward use during sleep only. NIPPV may be needed following hospital discharge and recovery.
Anesthesia/Pain	• Anesthesia consultation for all patients prior to surgery with general anesthesia • Evaluate airway carefully and plan for intubation difficulties given patients' physical limitations and potential abnormalities (e.g., decreased mouth opening, enlarged tongue) • Consider indirect visualization of the larynx including the possible fiberoptic intubation (i.e., fiber optic visualization is often necessary)	• Consider use of total IV anesthesia technique for induction and maintenance of general anesthesia • Avoid depolarizing muscle relaxants (i.e., succinylcholine) and non depolarizing muscle relaxants • If non depolarizing muscle relaxants used, titrate dose and continuously monitor neuromuscular function • Avoid inhaled anesthetics in DMD patients; these agents may be considered in SMA patients • Intubation without neuromuscular blockade is preferred when possible • Short acting opioids are suitable for intraoperative use in SMA patients	• Sedating analgesia may have additive effect on baseline respiratory insufficiency however postoperative pain control should not be compromised because of respiratory suppression concerns.[1,2] • Opiate-based analgesia in combination with acetaminophen and NSAID should be considered as part of routine post-procedural management with anticipation of providing appropriate NIV and cough assistance. • Patients may also benefit from benzodiazepines for muscle spasms and discomfort.
Nutrition	• Nutritional status assessment for all patients prior to surgery with goal to optimize nutrition and plan nutrition support during hospitalization. • Swallow evaluation may be helpful to determine ideal post operative feeding strategy • Pre-admit patients for total parenteral nutrition (TPN) if they require overnight enteral feeds. Patients with SMA have a fatty acid oxidation metabolic disorder as part of having SMA and do not tolerate prolong periods of time without nutrition.	• Consider continuing TPN infusion if started preoperatively. • Monitor glucose status intraoperatively to avoid hypoglycemia.	• Initiate bowel regime to avoid and treat constipation, with consideration or prokinetic GI medications as ileus is common • Any patient who cannot achieve adequate oral nutrition within 24-48 hours after surgery should receive enteral feeding with small-diameter nasoduodenal tube or parenteral nutrition • Consider gastric decompression with nasogastric tube in patients with GI dysmotility • If patient is receiving TPN, continue TPN until patient tolerates receiving at least 50% of goal enteral feed.
Cardiac	• Cardiac consultation for all DMD patients prior to surgery • For patients with history of symptoms suggestive of cardiac involvement or at risk for cardiac dysfunction, cardiac evaluation should be completed within 3-6 months prior to anesthesia.	• Close cardiac monitoring	• Monitor cardiac and fluid status postoperative • For DMD patients, obtain postoperative cardiac consultation • Use telemetry monitoring
Other	• Discuss goals of care, tracheostomy potential, prolonged dependency on mechanical ventilation and advance directives with patient/family	• .	• Consult Palliative care for goals of care planning, pain management considerations

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Additional Links

http://video.posna.org/media/The+Orthopedic+Management+Spinal+Muscular+AtrophyA+New+Treatments%E2%80%A6Next+Steps/1_0d4j9im6: POSNAcademy Webinar, The Orthopaedic Management of Spinal Muscular Atrophy: New Treatments…Next Steps, originally broadcasted in February 2019

https://www.curesma.org/: SMA resource for families and healthcare providers

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