

# Orthopedics

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## Introduction

Orthopedic problems and musculoskeletal deformities are common in patients with Spina Bifida. The prevalence of spinal deformities including scoliosis and kyphosis is proportionate to the severity of the neurologic lesion. Spinal deformities can be challenging to treat. Body casting that can be used in idiopathic early onset scoliosis may cause skin breakdown due to insensate skin. Bracing can be used, but there is limited data to support it. Surgical treatment carries high risks including infection, pseudarthrosis, and loss of mobility. Recent studies have questioned the effect of surgery on overall function.<sup>1-5</sup> Nevertheless, patients with early onset scoliosis or gibbus (kyphus) deformity present special challenges due to concerns about pulmonary function. Growing rod strategies such as spine to spine or rib to pelvis distraction are increasingly used for these patients, but the small size of the patient relative to the construct and poor soft tissue envelope remain challenges.<sup>6-8</sup> Further studies are needed to understand the risks and benefits of spinal surgery versus acceptance of the natural history of the spinal deformity.

Lower limb deformities can also be problematic and can affect function and gait. These can include contractures of the hip or knee or rotational deformities. Correction of rotational deformities, particularly external tibial torsion, has been shown to improve gait parameters on computerized gait analysis testing.<sup>9</sup> Release of contractures can also render the limb more amenable to bracing.

Hip subluxation and dislocation are common due to muscle imbalance, particularly in the patient with mid-lumbar lesion. In previous times, hip reduction surgery, including bone procedures and muscle transfers were commonly performed. A recent study suggests that hip reduction surgery is of questionable benefit in myelomeningocele while computerized gait analyses show that contracture, not subluxation, has a deleterious effect on ambulation.<sup>10</sup> Therefore, the use of hip reduction surgery has waned in recent decades, with the possible exception of individuals with low lumbar or sacral neurologic levels. For that reason, the present guidelines do not recommend routine surveillance of the hip or surgical treatment of hip subluxation/dislocation, although patients with a low lumbar or sacral lesion with unilateral dislocation could be considered an exception and must be treated on an individual basis.<sup>11-12</sup>

The non-plantigrade foot is a frequent problem in myelomeningocele. The feet typically manifest sensory impairments and consequently, skin breakdown can occur. Orthoses such as ankle-foot orthoses (AFOs) can support the foot in patients with motor impairments, but some foot deformities may preclude bracing. A variety of orthopedic strategies, both operative and non-operative, can be used to treat foot deformities. These include stretching, bracing, serial casting, and surgery. Surgery can include tendon releases and resections, tendon transfers, joint capsular releases, osteotomies, and fusions. In general, the younger, less rigid foot may respond to soft tissue procedures while the older or more rigid foot may also require osteotomy. A classic dictum is that fusions should be avoided when possible as they render the foot more rigid which can increase the risk of skin breakdown (clinical consensus). Nevertheless, some deformities may be sufficiently severe as to require salvage procedures such as talectomy,

subtalar arthrodesis<sup>14</sup> or triple arthrodesis.<sup>15</sup>

The proper timing for foot surgery is debatable, but a foot deformity that has become so severe as to be unbraceable in someone who still wants to pursue ambulation is certainly an appropriate indication. However, some surgeons may take a more proactive approach, performing tendon balancing surgery earlier in life to prevent bony deformity later. For example, a patient with an L4-level lesion with an unopposed anterior tibial tendon function will generally develop a calcaneus deformity. With time, weight bearing will be only on the calcaneus with no weight bearing through the forefoot. This can lead to calcaneal skin breakdown. Anterior tibial tendon transfer to the Achilles or merely anterior tibial tendon release done at an early age can improve or prevent this. If done later in life, a calcaneal osteotomy may be necessary.<sup>13,16</sup> Ultimately, the approach that is taken will be at the discretion of the surgeon and the family.

## Outcomes

### Primary

1. Maintain a stable and balanced spine.
2. Optimize pulmonary function and avoid restrictive pulmonary disease.
3. Optimize spinal growth.
4. Avoid or facilitate healing of sacral/ischial decubiti.

### Secondary

1. Maintain plantigrade feet.
2. Prevent skin breakdown.

### Tertiary

1. Preserve or improve gait efficiency.
2. Early identification and stabilization, or correction, of lower limb deformities.

## 0–11 months

### Clinical Questions

1. What are the consequences of early onset scoliosis, kyphosis and pulmonary insufficiency syndrome in patients with Spina Bifida?
2. Which foot deformities merit correction in the child 0-11 months old, and what is appropriate treatment?

### Guidelines

1. Perform neonatal kyphectomy, if required to facilitate skin closure.<sup>6</sup>
2. Orthopedic evaluations are recommended every three months in the first year of life. (clinical consensus)
3. Consider hip imaging using ultrasound in the infant and anteroposterior pelvis radiographs after 6 months in patients with low lumbar or sacral lesions. Consider using a rigid abduction orthosis to treat hip instability, but only in children with low lumbar and sacral deformities. (clinical consensus)
4. Ponseti casting or release is recommended for clubfoot or congenital vertical talus deformities.<sup>17-19</sup>
5. Perform spine evaluations by conducting a physical exam. Obtain scoliosis radiographs if a spinal deformity is suspected and monitor the spine for the progression of the deformity. In children who have not achieved sitting balance, perform radiographs in a supine position. Once sitting balance is achieved, perform spinal radiographs in a sitting position. (clinical consensus)
6. Consider bracing or casting when there is a documented progression of scoliosis.<sup>20</sup>

## 1-2 years 11 months

### Clinical Questions

1. What is the proper timing for correction of rotational deformities of the femur and/or tibia?
2. Are twister cables useful for rotational deformities?
3. What is the role of bracing or Mehta casting for early onset scoliosis?
4. Should gibbus deformity be treated surgically?
5. Is rib to pelvis distraction rather than kyphectomy the optimum treatment for gibbus deformity?

### Guidelines

1. Monitor the spine for development or progression of a deformity that may be due to a tethered cord or syrinx. Obtain anteroposterior and lateral scoliosis radiographs if a deformity is suspected on clinical exam. Perform radiographs in a sitting position if the patient is able to sit but not able to stand or in a standing position if the patient is able to stand. Repeat radiographs every one to two years if the deformity is present, depending on rate of progression. (clinical consensus)
2. Evaluate for neurologic changes or progression of scoliosis and discuss with neurosurgery specialists. (clinical consensus) (Neurosurgery Guidelines)
3. Initiate treatment for progressive early onset scoliosis that may involve casting<sup>21</sup> or bracing.
4. Consider tendon releases/transfers for unbalanced foot deformities such as the calcaneus foot<sup>22</sup> or equinovarus foot, if the foot is unbraceable, to facilitate orthotic management.
5. Consider twister cables for significant rotational deformities to facilitate ambulation until such time as surgical correction is appropriate.<sup>23</sup>
6. Surgical correction of rotational deformities of the tibia or femur is recommended only if they are limiting further motor development and causing difficulty with bracing. (clinical consensus)
7. Teach families about fractures and related precautions. (clinical consensus)

## 3-5 years 11 months

### Clinical Questions

1. Is bracing effective for early onset, non-congenital scoliosis?
2. Is rib-pelvis distraction versus the spine-based growing rod construct the optimal treatment for progressive scoliosis?

### Guidelines

1. Evaluate gait with careful attention to orthopedic deformities that render gait inefficient and preclude orthotic management. (clinical consensus)
2. Consider derotational osteotomy when rotational abnormality adversely impacts ambulation.<sup>24</sup>
3. Consider treating foot deformities with stretching, casting, bracing, soft tissue release or tendon transfers to facilitate orthotic management. (clinical consensus)
4. Evaluate the spine clinically and obtain scoliosis radiographs every one to two years if a progressive spinal deformity is suspected. Perform radiographs in a sitting position in children who can sit but not stand and in a standing position in children who can stand. (clinical consensus)
5. Work with neurosurgery specialists to determine whether a neurogenic cause of scoliosis progression is present. (clinical consensus) (Neurosurgery Guidelines)
6. Consider bracing for progressive, non-congenital scoliosis in the 25 to 50-degree range. (clinical consensus)

7. It is recommended that surgical treatment of scoliosis be reserved for a progressive deformity that is unresponsive to non-operative management. For example, when there is progression of the scoliosis in spite of bracing and after a neurosurgical cause, such as a tethered cord, has been ruled out. It is also recommended that management with growing rod surgery and fusionless technique should include spinal cord monitoring in patients with distal neurologic function.<sup>25</sup>
8. Consider surgical treatment of gibbus deformity for intractable skin breakdown or to free up the upper limbs for independent sitting.<sup>26</sup>
9. Teach children and families about fractures and related deformities. (clinical consensus)

## **6-12 years 11 months**

### **Clinical Questions**

1. Is bracing effective for early onset, non-congenital scoliosis?
2. Is rib-pelvis distraction versus the spine-based growing rod construct the optimal treatment for progressive scoliosis?

### **Guidelines**

1. Monitor gait, rotational deformities and foot position. (clinical consensus)
2. Consider correction of foot deformities to facilitate orthotic management with soft tissue release, tendon transfer and osteotomy, if necessary. It is recommended that fusion be avoided if possible. (clinical consensus)
3. Consider correction of tibial and femoral rotational deformities when they are interfering with gait and precluding orthotic management.<sup>24</sup>
4. Consider conducting computerized gait analysis, when available, in children with low lumbar or sacral level lesions who have atypical gait abnormalities. This information will be helpful when making decisions regarding surgery or bracing.<sup>3,16</sup>
5. Monitor for the development of scoliosis/kyphosis. (clinical consensus)
6. Obtain anteroposterior and lateral scoliosis radiographs every one to two years if deformity is suspected clinically. Do so more frequently in patients with progressive spinal deformity. Perform radiographs in a sitting position in those who can sit but not stand and in a standing position in patients who can stand. (clinical consensus)
7. It is recommended that surgical treatment of scoliosis be reserved for a progressive deformity that is unresponsive to non-operative management. An example is when the scoliosis has progressed in spite of bracing and after a neurosurgical cause, such as a tethered cord, has been ruled out. It is also recommended that management with growing rod surgery and fusionless technique should include spinal cord monitoring in children with distal neurologic function.<sup>14</sup> Growing rod surgery with sacral-pelvic fixation is effective in correcting deformity and achieving growth.<sup>17</sup>
8. Consider surgical treatment of gibbus deformity for intractable skin breakdown or to free up the upper limbs for independent sitting.<sup>15</sup> The current literature describes multiple techniques.<sup>26,28-32</sup>
9. Teach children and families about fractures and related precautions. (clinical consensus)

## **11-17 years 11 months**

### **Clinical Questions**

1. What is the impact of scoliosis/kyphosis on gait, sitting balance, and upper limb function?
2. What is the relationship between spinal deformity and skin breakdown?

3. Which patients benefit from spinal deformity surgery?
4. How can spinal deformity surgery be safely accomplished?
5. In lumbar scoliosis, how high must the fusion extend?

### **Guidelines**

1. Monitor for the development or progression of scoliosis clinically, with radiographs as necessary, if indicated by the physical exam. Perform radiographs in a sitting position in those who can sit but not stand and in a standing position in those who can stand. If the curve has progressed to an operative magnitude (50 degrees), discuss the risks and benefits of surgical treatment with the family. (clinical consensus)
2. Monitor for deterioration of gait and consider treatment of orthopedic deformities leading to deterioration such as hip and knee contracture or rotational deformities. Computerized gait analysis may be useful for decision-making in the case of children with low lumbar and sacral level lesions.<sup>11,16</sup>
3. Conduct a history and physical examination (with radiographs, if indicated) on an annual basis, unless greater frequency is indicated. (clinical consensus)

## **18+ years**

### **Clinical Questions**

1. What is the optimal orthopedic transition plan?
2. What degenerative issues can be expected for specific levels of function (e.g., knee arthrosis for mid-lumbar lesions with valgus thrust gait pattern) and what treatments can mitigate against these problems (e.g. knee, ankle, and foot orthosis (KAFO) or crutches for the above example)?

### **Guidelines**

1. Develop an orthopedic transition plan. (clinical consensus) (Mobility Guidelines).
2. Counsel the patient about potential orthopedic degenerative problems. Consider bracing across the knee, such as the use of a KAFO, for patients with coronal plane valgus knee stress, or adding forearm crutches to decrease coronal and transverse plane trunk motion.<sup>11</sup>
3. Counsel the patient about fractures and related precautions. (clinical consensus)

## **Research Gaps**

1. What are the consequences of early onset scoliosis, kyphosis, and pulmonary insufficiency syndrome in patients with Spina Bifida?
2. Is bracing effective in treating developmental (non-congenital) scoliosis in individuals with myelomeningocele?
3. Is Mehta-casting effective in non-congenital early onset scoliosis in myelomeningocele?
4. What is the impact of scoliosis/kyphosis on gait, sitting balance, or upper limb function?
5. Should gibbus deformity be treated surgically? What is the optimal age and surgical procedure for repairing gibbus deformities?
6. What is the relationship between spinal deformity and skin breakdown?
7. Which patients benefit from spinal deformity surgery?
8. Which patients with scoliosis will benefit from sacral-pelvic instrumentation, or one-stage versus two-stage operation? An evidence-based review concluded that spine surgery, if performed, should be through both an anterior and posterior approach.<sup>2</sup>
9. How high must the fusion extend in patients with lumbar scoliosis?

10. What is the proper timing for correction of rotational deformities of the femur and/or tibia?
11. What is the relationship between specific foot deformities and the development of skin breakdown? Does foot deformity surgery alter the risk of skin breakdown?
12. Is there an ideal, specific orthopedic transition plan?
13. Are twister cables useful for rotational deformities?
14. Which foot deformities merit correction in the child 0-11 months old, and what is appropriate treatment? What degenerative issues can be expected for specific levels of function (e.g., knee arthrosis for mid-lumbar lesions with valgus thrust gait pattern) and what treatments can mitigate against these problems (e.g. knee, ankle, and foot orthosis (KAFO) or crutches for the above example)?

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