INTRODUCTION

The past decade has seen a significant change in how orthopedic problems in children with spina bifida are approached. For example, instead of focusing on whether a patient’s hip is located or dislocated, more attention is placed on how the patient is functioning as well as the patient’s overall satisfaction. A shift has taken place emphasizing patient-oriented measures opposed to process measures such as measuring the degree of scoliosis correction. Gait analysis has significantly improved our understanding of the multiple factors that can affect the knee, which can help prevent knee dysfunction. Gait analysis has also improved our understanding of gait pathology and ability to measure outcomes. Foot deformities are generally treated with soft-tissue releases and osteotomies rather than arthrodesis to preserve motion. The goal of this article is to review the important advances made over the past 10 years in the management of spina bifida patient with musculoskeletal problems.

LOWER EXTREMITIES ISSUES AND GAIT ANALYSIS IN SPINA BIFIDA

Functional Neurosegmental Levels

The orthopedic management of patients with spina bifida consists primarily of preventing or correcting deformities to maximize mobility and independence while staying within the realistic expectations of the patient’s functional neurosegmental level. Treatment is primarily based on the patient’s functional neurosegmental level; however, it also requires a stable neurological status and knowledge of the natural history of spina bifida.

Walking capability is primarily affected by the neurosegmental motor level. However, a patient’s neurological level does not necessarily correlate to their ambulatory level. In a study by Bartonek et al. [2001], only 21 of 53 achieved expected ambulatory potential based on manual muscle testing systems to classify the neurological level in patients with spina bifida. However, these systems are not consistent and patients can be classified as L5 in one system and L4, for example, in another. The International Myelodysplasia Study Group has proposed a straightforward and easy-to-use system [Wright, 2001].

Walking capability is primarily affected by the neurosegmental motor level. However, a patient’s neurological level does not necessarily correlate to their ambulatory level. In a study by Bartonek et al. [2001], only 21 of 53 achieved expected ambulatory potential based on manual muscle testing. In fact, 22 of 53 patients were worse than expected and this was usually from poor balance, spasticity, and increased number of shunt revisions. Spasticity can occur in any patient with spina bifida and it has been shown to have a negative effect on the gait of patients with sacral to mid-lumbar spinal cord involvement.

Key Words: spina bifida; myelomeningocele; hip dislocation; valgus knee; foot and ankle deformity; bracing; gait analysis; scoliosis; kyphosis
dysraphisms. Spasticity and inadequate balance are negative determinants for patients to achieve ambulatory independence [Bartonek et al., 2005].

### Tethered Cord

The tethered cord syndrome is much better recognized and occurs in ~11% to 27% of patients with spina bifida. It can present with motor sensory changes in the lower extremities, spasticity, scoliosis, back and leg pain, and bladder changes [Michelson et al., 2004]. The natural history of untreated tethered cord syndrome in patients with spina bifida is poor. Phuong et al. [2002] found that 40 of 45 patients with spina bifida and tethered cord, who underwent bladder augmentation or contracture release, subsequently required additional surgical procedures because of complications of the tethered cord. More recently, Bowman et al. [2009] demonstrated a benefit of tethered cord release in a long-term study of 114 patients. They found that pain has the best response to surgical un tethering with 100% of children experiencing postop improvement. Seventy percent of patients showed improved lower extremity muscle strength compared to preoperative testing and gait was also improved. Spasticity improved in two-thirds of the patients and lower extremity contractures were stabilized in 78%. Urologically, 64% of patients showed improvements on postop bladder evaluation [Bowman et al., 2009].

### Hip Disorders

The management of hip disorders has undergone a radical change over the years. Using gait analysis, Gabrieli et al. [2003] reported that gait symmetry corresponds best to the absence of hip contractures and has no relation to the presence of hip dislocation. They evaluated 20 patients with lower lumbar myelomeningocele using gait analysis. All were community ambulators with solid AFOs and crutches and a unilateral hip dislocation or subluxation but no surgical treatment. Seventy percent of patients who had no contracture walked with a symmetrical pattern, whereas only 20% of patients who had a unilateral hip flexion and/or adduction contracture walked with a symmetrical pattern. The authors conclude that reduction of the hip is unnecessary and recommended treatment of the contracture alone [Gabrieli et al., 2003].

Correll and Gabler [2000] found similar results to Gabrieli et al. [2003] in a report of 55 patients with spina bifida and hip contracture treated with soft tissue releases only. They found that a subluxated or dislocated hip did not influence the final outcome and most of the patients obtained a great advantage from the operation. Hip flexion contractures have been correlated to the degree of lumbar spine lordosis in spina bifida, but it is not known whether extension osteotomies would benefit the patients [Glard et al., 2005]. Although the trend has been toward less aggressive hip surgery, an interesting report by Lorenté-Molto and Garrido [2005] suggests that the posterior-lateral transfer of iliopsoas (Sharrard technique) was beneficial in a group of 29 children with L3 level spina bifida. Swaroop and Dias [2009] recently summarized the literature regarding hip management in spina bifida and concluded that although surgical treatment may allow reduction of the dislocated hip, this must be weighed in terms of the potential for complications and functional decline.

### Gait analysis has significantly increased our ability to evaluate knee and leg function and to better understand factors which affect the knee.

### Knee Issues in Spina Bifida

Knee pain is common in ambulatory persons with spina bifida and may be a significant reason for patients choosing not to walk. Gait analysis has significantly increased our ability to evaluate knee and leg function and to better understand factors which affect the knee. In the stance phase of gait, the knee is subject to the forces of the upper body and the ground reaction force, coming through the foot which can force the knee into a valgus position. This can produce excessive medial stress on the knee which can be measured as an adductor moment and commonly called coronal plane valgus knee stress. Several groups have investigated knee function in spina bifida focusing on coronal plane knee valgus stress. A number of factors have been identified as contributing factors to abnormal knee moment and decrease knee pain.

Weak hip abductor muscles have been correlated with development of abnormal knee motion and valgus knees because of the significant lateral sway when walking or using AFOs. The use of crutches with AFOs or KAFOs has also been recommended to protect the knee. Distal tibial derotational osteotomies have been reported for patients with excessive ETT. However, the operation does not result in complete normalization of their knee moments [Dunteman et al., 2000]. The cause of coronal plane valgus stress is multifactorial and influenced by increased lateral trunk sway, weak hip abductors muscles, internal hip rotation in combination with increased knee flexion, ETT, and valgus foot deformity [Gupta et al., 2005].

### Foot and Ankle in Spina Bifida

Foot and ankle deformities are very common in spina bifida with equinus contracture, clubfeet, and calcaneal deformities (excessive dorsiflexion) being the most common [Frischut et al., 2000]. The widespread application of the Ponseti method for the treatment of idiopathic clubfeet has been used in spina bifida patients with clubfeet.

Gerlach et al. [2009] reported an early relapse of the deformity in 68% of patients. Twenty-five percent eventually required extensive soft tissue releases. Future studies using the Ponseti technique may help define proper patient selection and its role in spina bifida.

Flynn et al. [2004] reported on 45 children with myelomeningocele who were treated with radical posterior remidal release without internal fixation and found that 61% had good results, 26% had fair results, and 13%
had poor results. Recurrent clubfeet are difficult to treat, but the authors of this article had very satisfactory results from using a takedown with calcaneo-cuboid fusion for recurrent clubfeet in spina bifida. This is a very straightforward operation that does not require the use of an external fixator [Dias et al., 1987].

Preservation of motion in treating foot deformities with osteotomies opposed to arthrodesis of joints has gained wider acceptance. Severe hindfoot valgus can be treated by medial displacement osteotomy of the calcaneus in children with spina bifida. Trotosian and Dias [2000] performed a retrospective review of 27 patients with severe hindfoot valgus and found that 82% of the patients were completely satisfied and most had complete correction of the hindfoot deformity in a group of 27 patients with 38 affected feet who have been followed for an average of greater than five years after surgery. Ponce and Kirkham [2009] reported their experience with the Grice (subtalar joint) arthrodesis in the treatment of valgus feet on 23 patients (35 feet). Interestingly, they report that ankle valgus worsened during follow-up but not significantly. The visual analog scale at final follow-up improved significantly and 83% were satisfied with the surgery. Valgus deformity can also occur at the ankle joint and can be easily treated with guided growth principles in a growing child, which allows for gradual correction of the deformity by tethering the medial side of the growth plate [Davids et al., 1997; Stevens et al., 1997].

Calcaneus deformity occurs in patients with unopposed anterior tibialis activity. Patients develop excessive dorsiflexion of the ankle, which puts excessive pressure on this area with subsequent risk of skin breakdown. Controversy exists in the literature as to whether the anterior tibial tendon should be released or transferred. Park et al. [2008] have shown that the posterior transfer of the anterior tibialis tendon with concurrent procedures to balance the muscular forces in the foot was successful in balancing the foot and improving pressure distribution under the foot. However, less improvement was noted in patients with increased pelvic rotation before surgery. These patients also had decreased knee extension in stance phase and increased hip abduction and pelvic obliquity both before and after surgery. Wenz et al. [2008] performed tendon transfers of the tibialis anterior, tibialis posterior, peroneus brevis and longus, and extensor digitorum and hallucis longus to the Achilles tendon, in addition to a triple arthrodesis (fusing the three joints about the hindfoot) to address a long standing calcaneal deformity. As noted above, close scrutiny of any foot arthrodesis with long-term follow-up is warranted, due to concerns with increased skin breakdown and adjacent joint arthritis following an arthrodesis.

Gait Analysis and Orthotics

Over the past decade, especially with the advent of gait analysis, the importance of hip abductor strength has become more appreciated. Ounpuu et al. [2000] demonstrated a correlation between hip abductor strength and the presence of abnormal knee moments (valgus stress). Guttierrez et al. [2005] identified hip abductor weakness as the one muscle most important in changing gait kinetics. Shoennakers et al. [2009] found that patients with myelomeningocele can have decreased 6-min walking distance and decreased aerobic capacity associated with weak hip abductors. They suggested that independent ambulating children would benefit from endurance training and muscle strengthening. Even at a sacral level, the muscle strength of the hip abductors and ankle dorsal-flexor muscles was strongly associated with ambulation in two groups of patients with myelomeningocele versus lipomeningocele [Shoennakers et al., 2004]. These studies suggest that exercise physiology may play an important role in future research studies.

Energy consumption and gait have been evaluated extensively. Thomas et al. [2001] evaluated oxygen consumption, energy cost, and velocity over time in 23 children with myelomeningocele to determine whether differences exist in children with hip, knee, ankle, foot orthoses (HKAFOs) versus reciprocating gait orthoses (RGOs). Children using HKAFOs had similar oxygen costs as children using RGOs while achieving a faster velocity. The authors found that upright ambulation may progress from ambulation with an RGO when the child’s upper extremity strength-to-mass ratio is low, to an HKFO when upper extremity strength improves and velocity or keeping up with peers is of concern [Thomas et al., 2001]. Moore et al. [2001] compared the energy cost of walking in low lumbar myelomeningocele patients between a reciprocal gait versus a swing-through gait pattern. They found that a swing-through gait pattern proved to be the more efficient walking pattern in this group of subjects with myelomeningocele.

Bare et al. [2001] evaluated 14 independent ambulating children with myelomeningocele and found that pelvic obliquity demonstrated the strongest relationship with oxygen cost, suggesting that ultimately, hip abductor strength may play a key role in energy demands during gait. Patients tended to walk with a decreased self-selected walking velocity in which many of these children are comfortable and may be predicated on an optimal center of mass moment that approximates the magnitude observed in normal gait. Therefore, patients may self-select walking speed to control their vertical and horizontal center of mass excursions to conserve energy [Bare et al., 2001]. Finally, Galli [2000] found that the use of AFOs in children with myelomeningocele leads to an improvement in gait and reduced energy consumption.

RGOs are recognized mobility aid for lower thoracic-upper lumbar paraplegia but are expensive and not all patients utilize them despite meeting prescription criteria. The Alyn Rehabilitation Hospital reported a novel program in which every patient who is a candidate for an RGO first undergoes an evaluation in a used but adjusted RGO. Only those who achieve a minimum of ten strides on repeated occasions receive their own RGO. Seventy percent were successful which correlated inversely with the neurological level and correlated directly with parental cooperation. [Katz-Leurer et al., 2004]

The use of carbon-fiber braces are important advances in the orthotic management of children with spina bifida. Two recent articles have demonstrated these outcomes. Wolf et al. [2007] found that the use of the carbon-fiber spring orthosis significantly increased the energy return during the third rocker (during the stance phase of gait) simulating the natural push-off action. Bartonek et al. [2007] found that carbon-fiber spring orthoses enhanced gait function by improving ankle plantar flexion moment, ankle positive work, and stride length. However, the carbon fiber spring orthosis did not suit all participants.

SPINAL DEFORMITIES IN SPINA BIFIDA

Associated Spinal Cord Abnormalities

The treatment of spinal deformities continues to be challenging despite the numerous advances in the past...
10 years. These complex three-dimensional deformities include both scoliosis and kyphosis, often occurring together. They are complex in that they have an early onset and rapid progression; have an absence of normal posterior muscle, bone, and skin; and have associated congenital spine anomalies. The incidence of spinal deformity in spina bifida ranges from 50% to 90% [Trivedi et al., 2009; Akbar et al., 2009] and is correlated with the individual’s motor or neurological level. Individuals with thoracic level myelomeningocele have the highest incidence followed by high lumbar, and individuals with low lumbosacral level have the lowest incidence of spinal deformity.

Scoliosis in Spina Bifida

The treatment goals for scoliosis in patients with spina bifida include preventing progression of the deformity, achieving a solid fusion, maximizing functional independence, increase sitting tolerance, and achieve a level pelvis with a balanced spine in both coronal and sagittal planes. The risk of deformity progression is associated with the patient’s neurological level, the last intact laminar arch (LILA) and the patient’s ambulatory status. In their study of 141 patients, Trivedi et al. [2002] reported a prevalence of scoliosis in 52%. Scoliosis developed before the age of nine in 43 patients. The clinical motor level, ambulatory status, and last intact laminar arch were all found to be predictive factors for the development of scoliosis. Glard et al. [2007] evaluated the neurological classification in spina bifida as a spine deformity predictor. The authors noted that patients in group 1 (L5 or below) was a predictor for the absence of spinal deformity. Patients in group 3 (L1-2) or 4 (T12 and above) was predictive for spinal deformity, and group 4 (T12 and above) was a diagnostic for developing a kyphosis.

The relative indications for operative treatment include scoliosis greater than 50°, children unable to be managed in a brace, and children older than 10–12 years of age to maximize adult spinal height. The benefits of surgery always need to be weighed against the potential risks and complications. Several studies have highlighted the potential complications. Geiger et al. [1999] evaluated their complications in a cohort of 77 patients with myelomeningocele and scoliosis. Complications were analyzed by instrumentation problems, pseudarthrosis, infections, anesthetic, and neurological complications. Instrumentation problems were seen in 29% of their patients. Pseudarthrosis was dependent on the surgical technique utilized, and the extent of the fusion including the pelvis. Patients just with hardware problems had a mean loss of correction of 49% compared with 13% in other patients. Early post-operative shunt failure was found in four cases, and three late presentations of shunt failure occurred after one year [Geiger et al., 1999].

A multicenter study assessed the risks for deep wound infections following neuromuscular scoliosis surgery [Spohr et al., 2000]. Deep wound infections developed in 16 patients with myelomeningocele in this retrospective review of 210 patients with myelomeningocele and cerebral palsy (CP). Of the 10 risk factors studied, the authors found that only the degree of cognitive impairment and the use of allograft were significant. Infections commonly were polymicrobial and caused by gram negative organisms. The authors also reported that patients in whom infection develops are at increased risk for developing a nonunion of their spine fusion [Spohr et al., 2000]. Deep wound infections were also noted to be a serious problem in other reported series in this patient population [Banit et al., 2001; Benson et al., 1998; Stella et al., 1998].

The use of anterior instrumentation and fusion in selected patients and segmental pedicle screw fixation are important advances in the surgical planning and techniques. Casillas et al. [2003] evaluated 21 patients who had anterior spine fusion with anterior instrumentation alone. The majority of these patients had myelomeningocele. Complications noted by the authors included progression of the curve adjacent to the instrumented segment, and one infection that required irrigation and debridement of the wound. In selected patients, anterior arthrodesis and instrumentation spanning a short segment of the spine can be an effective procedure for scoliosis, even with an associated pelvic obliquity. The potential advantages of this approach are the preservation of motion segments, and maintaining normal myelocontour.

In a similar study, Spohr et al. [1999] evaluated the efficacy of anterior only fusion in 14 patients with myelomeningocele. The indications for surgery included thoracolumbar scoliosis greater than 45°, a compensatory thoracic curve of less than 40°, and the absence of a significant junctional kyphosis. Thirteen patients had prior neurosurgical intervention for a tethered cord, ACM, or syringomyelia. Anterior instrumentation spanning an average of seven levels, the primary curve was corrected a mean 57% at follow-up of 40 months. The associated pelvic obliquity corrected from a mean of 16° to 9°. Loss of correction, similar to the above study, occurred in the segments of the spine proximal and distal to the instrumented levels. Two patients, both with
syringomyelia, required additional surgery due to proximal decompensation of the deformity. The authors noted good results in five patients, fair in four, and poor in five. Neurological deterioration occurred in two patients with preoperative curves greater than 75°. There were no deep wound infections, and only one superficial wound infection. The authors concluded that the intended goals of an anterior only approach in spina bifida patients; decreasing the infection rate and achieving acceptable correction of the scoliosis and pelvic obliquity could be attained for selected patients. The recommended indications for selected patients include a thoracolumbar scoliosis less than 75°, no evidence of syringomyelia, compensatory curves less than 40°, and no increased kyphosis [Sponseller et al., 1999].

Pedicle screws offer the ability to achieve posterior segmental fixation in the myelodysplastic spine that previously was difficult to obtain with other forms of fixation such as hooks and wires that required the presence of an intact laminae. The limitations of pedicle screw fixation are the abnormal pedicles in this patient population, which are often small, dysplastic, and mal-oriented [Guille et al., 2006]. Parisini et al. [2002] evaluated the use of pedicle screw fixation to improve results in patients with myelomeningocele. The authors compared three groups of patients defined by different treatment approaches. Staged anterior and posterior spine fusion and instrumentation yielded good correction and stability. Deformity correction using a posterior only approach with pedicle screw segmental fixation and posterior fusion, even for severe deformities, was found to be effective. This confirmed the findings from an earlier report that noted additional benefit of pedicle screw fixation including correction and preservation of lumbar lordosis [Rodgers et al., 1997].

Consensus exists whether or not to extend the posterior instrumentation and fusion to the pelvis to address the scoliosis deformity and associated pelvic obliquity. Achieving a solid fusion across the lumbosacral joint can be difficult with absent posterior elements, and instrumentation failure is frequently noted. Even when a fusion is obtained, it may limit the ability of ambulatory patients to walk [Akbar et al., 2009]. Wild et al. [2001] evaluated the results and functional outcome in 11 patients who underwent staged anterior and posterior fusion and instrumentation without including the sacrum. The authors noted spontaneous correction of the pelvic obliquity when the scoliosis deformity was addressed without including the sacrum. The authors concluded that sparing lumbar segments provided improved mobility. Akbar et al. [2009] recommended not extending the fusion and instrumentation to the pelvis if the lumbar curve can be corrected to <20° and the pelvic obliquity to <15°. However, the authors noted that patients often present at advanced stages of their scoliosis, which makes the treatment more challenging and often demands extending the fusion to the pelvis.

Several recent articles have addressed the need for single or staged anterior and posterior spine fusion in myelomeningocele. Parsch et al. [2001] evaluated their experience in 54 patients with scoliosis secondary to myelomeningocele. They reviewed three different surgical techniques used in treating this difficult problem. The third group with anterior and posterior instrumentation demonstrated better long-term correction compared to Group I. Stella et al. [1998] evaluated their experience in 20 patients with severe scoliosis due to myelomeningocele. Nineteen of the patients underwent combined anterior and posterior fusion with instrumentation, seven patients were treated with posterior arthrodesis alone with instrumentation, and three by anterior arthrodesis with instrumentation. The fusion was extended to the sacrum in 15 patients. Combined anterior and posterior instrumentation and fusion provided the best correction of deformity, and decreased the rate of pseudarthrosis to 14%. Banit et al. [2001] reviewed their experience with posterior spine fusion in 50 patients with myelomeningocele. They reported a complication rate of 48%, deep wound infection rate of 8%, and a pseudarthrosis rate of 16%. Despite their description of modern segmental instrumentation decreasing the pseudarthrosis rates for posterior spine fusion alone, the authors recommend the combined anterior and posterior approach for scoliosis in the myelodysplastic patient [Banit et al., 2001].

**Kyphosis in Spina Bifida**

The incidence and pathophysiology of rigid kyphotic deformities have been described in two recent review papers [Guille et al., 2006; Akbar et al., 2009]. The effect of spinal cord transection or cordotomy associated with kyphectomy has been recently evaluated. Proponents of cordotomy assert the advantage of improved correction when combined with a kyphectomy. Ko et al. [2007] reviewed their institution’s experience in nine children with myelomeningocele treated with a combined spinal cord transection and kyphoscoliosis correction, with an emphasis on evaluating the changes in cerebrospinal fluid (CSF) dynamics that may occur after cordotomy and ligation of the distal CSF circulation. Eight of the children had postoperative complications (89%) including wound infection or skin breakdown. One patient had a CSF leak, and two children required revision of their VP shunts within six weeks of surgery. The authors contributed this to alteration of CSF dynamics resulting from loss of CSF absorption and flow buffering capacity below the level of the spinal cord transection. A mean correction of the kyphotic deformity of 81° was achieved with this combined technique. Lalonde and Jarvis [1999] evaluated the effect of spinal cord transection on bladder function for correction of congenital kyphosis and myelomeningocele. Bladder function below and after the procedure was assessed clinically and quantitatively by urodynamic studies in thirteen patients with a mean age at time of surgery of 8.9 years. Only one patient exhibited a decrease in bladder function after the operation. Eight of the patients who had urodynamic assessment recorded improvement of bladder capacity and compliance, and five demonstrated an increase in urethral pressure.

Debate also continues related to the timing of surgery for rigid kyphotic deformities and the techniques used for correction. Neonatal kyphectomy has been associated with a high rate of recurrence. In a retrospective study, Crawford et al. [2003] evaluated a group of 11 patients who underwent neonatal kyphectomy and dural sac closure. For this cohort of patients, the mean initial correction was 77°, with a loss of correction at follow-up of 55°. The authors reported successful closure on all nine infants, and no serious complications were noted. One patient required a repeat kyphectomy at nine years of age. The authors concluded that neonatal kyphectomy performed at time of dural sac closure is a safe procedure and provides excellent initial correction. Recurrence of the deformity is expected, but the resultant kyphosis that
occurs in longer and less angulated, which theoretically should be less technically demanding.

Nolden et al. [2002] evaluated their experience with a subtraction kyphectomy technique with posterior instrumentation in 11 children with myelomeningocele. The authors described subtraction kyphectomy as a technique that removes the cancellous bone (decellulation) of the apical vertebra to correct the rigid deformity without excising the vertebrae. This allowed the authors to preserve the thecal sac, and limit arthrodesis of the spine with pedicle screw instrumentation in young patients. The mean age at index procedure was six years, and the mean preoperative kyphosis was 88°.

The deformity was corrected down to a mean of 3° of lordosis, with an average loss of correction of 24° at two-year follow-up. The authors reported no acute onset hydrocephalus, no deaths, vascular complications, or deep wound infections with this technique. Conclusions drawn from this study were that this technique of subtraction decancellation vertebrectomy with preservation of the dural sac is safe, and facilitates correction and stabilization of kyphosis in young patients with myelomeningocele.

Thomsen et al. [2000] reviewed their experience with the Fackler technique or insertion of rods through the S1 foramen [Warner et al., 1993]. Nine patients had a mean preoperative kyphosis of 152°. Using this technique, the authors achieved a 67% correction. The proximal levels were left unfused for future growth. Two complications with the distal fixation were described. The authors stated that the technique provided an effective means of distal fixation to correct the angulation of the pelvis, which should be limited to patients weighing less than 30 kilograms [Thomsen et al., 2000].

In an updated study from the same institution, Akbar et al. [2006] retrospectively reviewed 24 consecutive patients who underwent the Fackler technique. The average preoperative lumbar kyphosis was 124°. The authors achieved an average correction down to 43° after surgery. In a subsequent study, Akbar et al. [2009] further reviewed their group of 28 patients evaluating the Fackler technique. Long-term complications secondary to implant failure were noted in nine patients, with revision surgery required in five. Biomechanical analysis demonstrated a correlation between implant failure and remaining deformity after the surgical procedure. In both studies, the authors concluded that surgery should always be performed with the intention to establish a normal sagittal profile to minimize the risk of implant failure [Akbar et al., 2006; Akbar et al., 2009].

Odent et al. [2004] evaluated nine patients who underwent a two-stage procedure for rigid congenital kyphotic deformities. The initial stage included a posterior kyphectomy with modified Dunn-McCarthy fixation (a long S-shaped rod contoured over the sacral alae and buttressing the anterior sacrum) and using lumbar pedicle screw fixation. The anterior second stage was completed several weeks later placing an inlay strut graft from typically from the T10-S1 levels. The patients had a mean correction from 110° to 15°. No instrumentation failures, loss of correction or pseudarthrosis were noted. Complications consisted on one intra-operative cardiac arrest that was reversible, one wound necrosis, and one deep venous thrombosis. This two stage technique was thought to enhance the biomechanical and biological anterior fusion mass, and prevent late instrumentation failure and subsequent loss of correction.

In a long-term outcome study, Niall et al. [2004] evaluated the results of kyphectomy in 24 children with myelomeningocele at ten year follow-up. The authors reported a high complication rate including delayed wound healing and late skin breakdown with exposure of instrumentation. Additional surgery to remove protruding hardware was required in 18 patients. Posterior instrumentation with fixation to the pelvis provided better stability on follow-up than other methods. The authors judged the long-term clinical and radiographic outcome to be excellent despite the prolonged morbidity in the majority of patients in the study.

Functional Outcomes in Spine Surgery

Several recent studies have examined the functional outcomes of patients following the operative treatment of spinal deformities in spina bifida. There has been a shift from emphasizing process measures such as measuring the degree of scoliosis correction to patient-oriented measures. To evaluate physical disability in children with scoliosis and myelomeningocele Wai et al. [2005] developed a valid and reliable instrument entitled the Spina Bifida Spine Questionnaire. Another study from the same institution evaluated the relationship of spinal deformity in myelomeningocele to physical function and self-perception [Wai et al., 2005]. Spinal deformity was determined by the degree of scoliosis, pelvic obliquity, and coronal sitting balance. Physical function was measured using the Activities Scale for Kids (ASK). Coronal balance was significantly related to only one measure of physical function. Using the ASK instrument, no relationship was observed between spinal deformity and global physical function. The authors concluded that indications for surgery in children with scoliosis and spina bifida should be well defined, and that the potential benefits of surgery in the short term may only improve sitting balance.

Schoenmakers et al. [2005] evaluated the effect of spine fusion on ambulation and functional abilities in children with spina bifida. Ten children were prospectively followed after spine surgery at a mean age of 9 years. Despite improvement in the primary curve after surgery, ambulation became difficult in three of the four patients who had ambulated preoperatively. This was particularly noted in exercise ambulators. Increased caregiver assistance is required for self-care and mobility within the first six months after surgery. In their cohort of patients, the authors noted that it took about one year to return to their presurgery levels, and only minimal improvement is observed afterwards.

Another major trend in orthopedic research is the focus on evidence-based studies. Sackett et al. [1996] defined evidence based medicine as the "conscientious, explicit, judicious use of current best evidence in making decisions about the care of individual patients" [p. 71]. Levels of evidence and grades of recommendation have been described to evaluate the current literature and may guide treatment decisions [Wright, 2007].

Two articles highlight the relatively poor level of evidence available on studies examining spinal deformities and spina bifida. Mercado et al. [2007] performed a systematic literature review regarding the quality of life in patients with different types of neuromuscular scoliosis who underwent a spine fusion. A grade C recommendation for spine fusion was given for both cerebral palsy and muscular dystrophy in improving quality of life. This is based on consistent Levels 4 and 5 studies, which "may" guide treatment decisions. The authors noted that spine fusion does not improve the quality of life in spina
bifida, which was given a grade C recommendation. Wright [in press] appraised the literature on the treatment of scoliosis and spina bifida, and found that the benefits of surgery are uncertain. This was given a Grade I, which is based on inconsistent or insufficient, evidence, and provides no guidance for treatment decisions. A Grade B recommendation is given for combined anterior and posterior procedures if spine surgery is considered. And the literature suggests (Level of Evidence I) that an all pedicle screw approach for posterior segmental instrumentation may be effective [Wright, in press]. These studies also strongly suggest that future studies should be prospectively designed, randomized or comparative, to improve the levels of evidence (I or II) for the evaluation and treatment of spinal deformities in spina bifida.

Future Research Directions
Despite the important advances made over the past decade in the orthopedic management of spina bifida, future research is needed on several fronts. Exercise physiology in spina bifida should focus on methods to increase muscle strength, especially the hip abductor muscles which are so important for efficient ambulation. Improvements in brace construction and design with stronger and lighter energy-storing materials (such as the newer carbon fiber orthotics) can result in improved ambulation. Oxygen consumption studies in conjunction with gait/motion analysis will be vital in objectively quantifying these improvements in orthotic design and outcomes after surgical procedures. Expanded use of functional outcome measures and evidenced based studies in spina bifida will help continue to refine the indications for operative intervention for lower extremity and spinal deformities.

REFERENCES


