Chapter

Orthopedic Approach to Spina Bifida

Roselle C. Okubo, Claudio Silveri and Ana C. Belzarena

Abstract

Spina bifida is a common nervous system malformation and it encompasses a wide array of presentations with diverse orthopedic challenges. Manifestations of this disease can include dislocates hips, joint contractures, spine deformity such as scoliosis or kyphosis, clubfeet and limb rotational deformities. Additionally, many of these patients are non-ambulatory and prone to osteoporosis induced pathological fractures. The care of spina bifida patients is a challenging one, requiring many health care professionals from different areas to be working in conjunction. Nowadays, spina bifida patients live longer due to advances in health care and improving the quality of life of these patients is paramount.

Keywords: spina bifida, myelomeningocele, orthopedic surgery

1. Introduction

Spina bifida is the most common nervous system malformation. This complex disease can be considered as a group of congenital defects caused by a failure in the closure of the neural tube at the fourth week of the embryonic phase [1]. The true incidence may vary from country to country but overall is at 0.5 per 1000 births [2]. Additionally, gender prevalence is more in girls than in boys, but again it varies geographically [3]. There are mainly two categories of spina bifida, open and closed ones. The open types which include meningocele and meningomyelocele have neural tissue exposed and are more severe in terms of symptoms and prognosis [4]. Closed spina bifida or occulta, has no neural tissue exposed and includes from lipomeningocele to just a sinus tract [5]. Majority of these neural tube defects are located at lower levels of the spine, mostly in the lumbar and sacral levels [6]. These defects can be diagnosed prenatally with ultrasound imaging or maternal alpha-feto-protein levels measured on the mother's serum. Patients with spina bifida can often present with neurological deficits, motor or sensory and orthopedic conditions such as joint contractures, spine deformity, clubfeet and hip dislocations among others. The degree of the deficit and the orthopedic presentation are related to the spine level where the defect is present [4].

2. Non-orthopedic health conditions

Besides the orthopedic associated conditions, these patients can present with several other health problems. The mortality of these patients has decreased throughout the years with enhanced medical care, thus now more attention is driven

at improving these patients' quality of life [7]. Intellectual disability is present only in approximately 20% of the patients and is usually the consequence of hydrocephalus [8]. Patients usually present with bladder and/or bowel incontinence, renal failure, propensity to infections and skin ulcers due to skin insensitivity, hydrocephalus, tethered cord and Arnold Chiari II type of malformation [9]. One in three of these patients will be allergic to latex, some having anaphylactic reactions. This is thought to be the consequence of repeated surgical and medical procedures, thus the importance of avoiding latex material since the beginning of care [9].

3. Pathologic fractures

Due to the lack of ambulation, physical exercise and axial bone load spina bifida patients can present with osteoporosis and osteoporosis induced fractures [10]. The

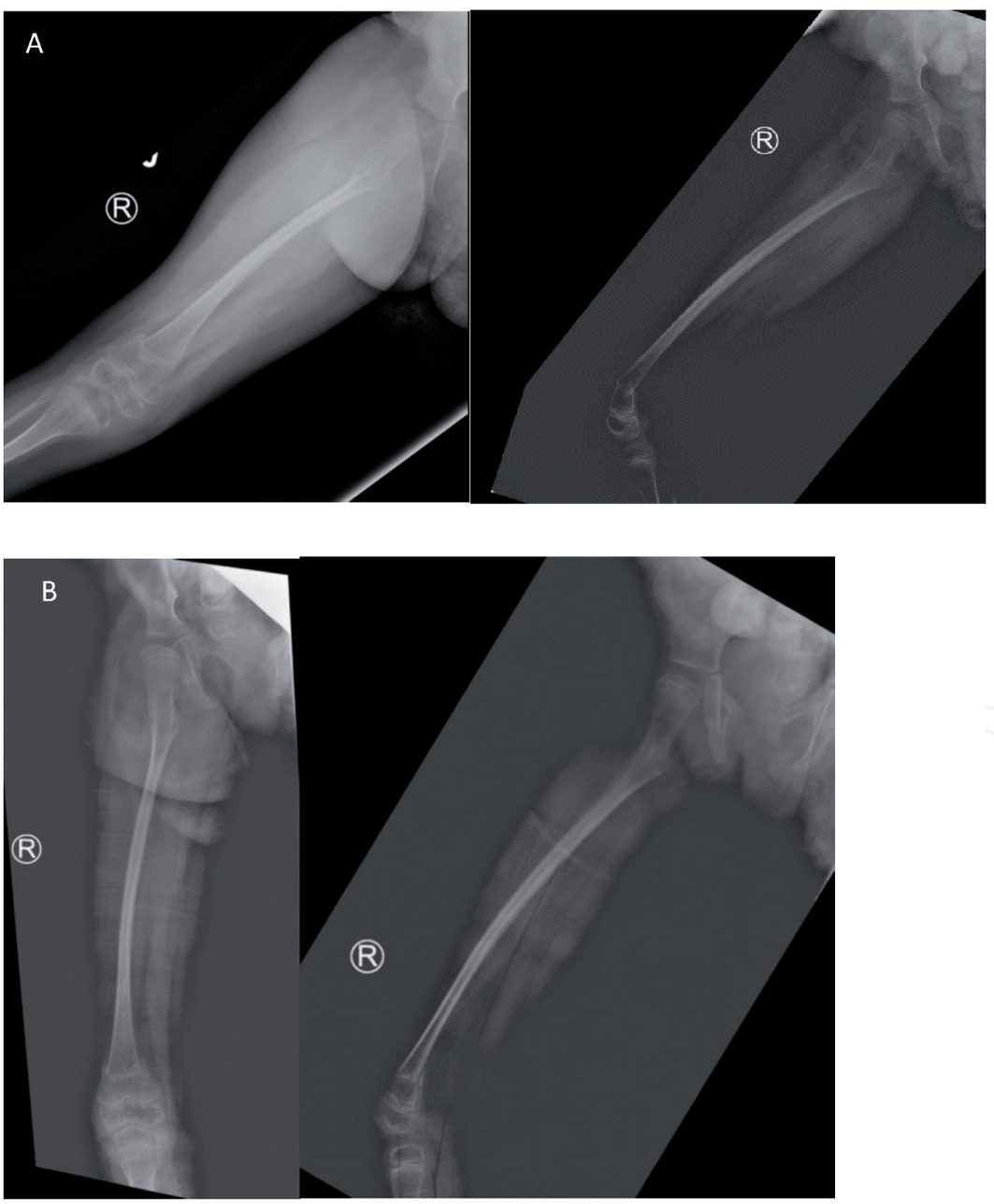

Figure 1.
Right distal femur fracture in a myelomeningocele patient without an obvious traumatic mechanism (A) and radiographic images of post-reduction and casting (B).

fractures usually occur below the neurological level of the defect and the incidence ranges from 11 to 30% [11]. The fracture mechanism is usually pathologic, these fractures usually being caused by minor trauma or even spontaneously [12]. Since many of these patients may have a fractured bone without an obvious trauma mechanism it can be difficult to diagnose these fractures. Patients usually present with a swollen, warm extremity with associated redness, and this should prompt obtaining a radiographic imaging study [13]. The caring orthopedist should be aware not to confuse these symptoms with an infection. The fractures are common the higher the level of the neural defect, in the distal femur or around the hip in patients from 3 to 7 years old (**Figure 1**) [14]. Treatment is usually non-surgical and involves immobilization in a cast. Prolonged immobilization in the cast should be minimized since this also will make osteopenia worse [15]. Patients should be assessed for bone density with dual-energy X-ray absorptiometry (DEXA scan) exams and calcium and vitamin D levels should be assessed and replaced if necessary, by the pediatrician. Weight bearing and physical exercise should be encouraged as appropriate [16].

4. Spine care

Besides the posterior element defect in the spine, spina bifida patients also present with severe congenital deformity and contractures of the spine. These deformities can pose a restriction to everyday activities as well as pulmonary function [17]. A third of the patients will have scoliosis, which is usually of an early onset and has a tendency to progress and cause pelvic obliquity [18]. Scoliosis has different causes in these patients such as muscle imbalance or primary malformations like hemivertebra and vertebral fusions. Kyphosis may also be present in approximately 15% of the patients (**Figure 2**). Is usually progressive and mostly located in the lumbar region [19]. The deformity can be so severe to cause skin breakdown at the level of the deformity (**Figure 3**). Surgery is necessary to correct the deformity and is not free of complications in these patients. Usually there is no role for bracing spine deformity in these patients and the skin insensitivity can predispose to skin ulcers and infection. Surgical correction is indicated in patients with progressing curves who are good candidates for surgery. Posterior fixation is the most common procedure performed but other options such as an anterior fusion or combined ones are used as well when appropriate. In patients with pelvic obliquity the fixation should be extended until the pelvis level, this is particularly important in non-ambulatory patients (**Figure 4**) [20]. Surgery can be associated with higher risks of infection, anesthesia complications, bleeding, non-union, hardware failure, loss

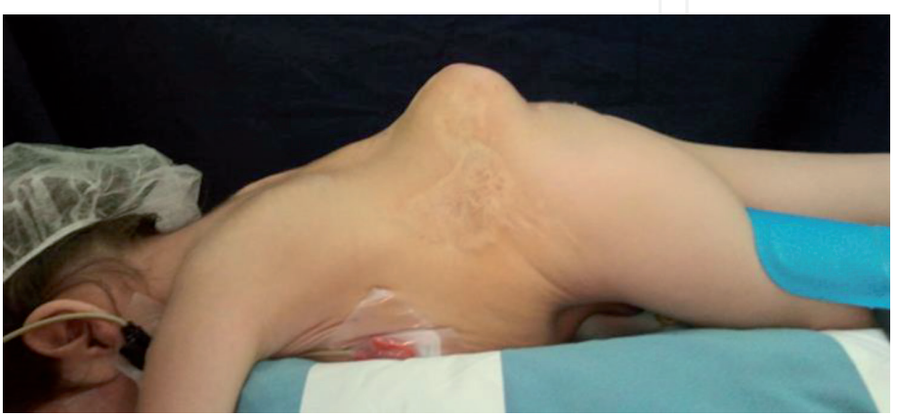

Figure 2.
Myelomeningocele patient with marked lumbar kyphosis.

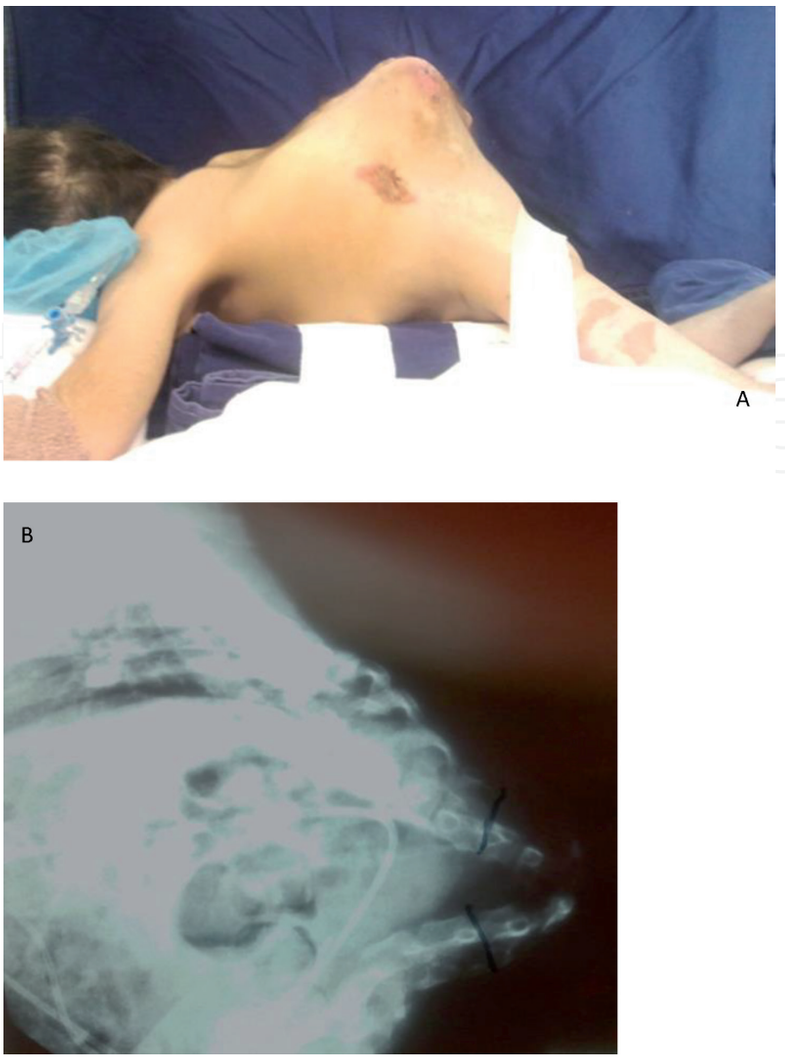

Figure 3.
Myelomeningocele patient with marked kyphosis with skin breakdown at the level of the deformity (A) and accompanying radiographic images of the deformity (B).

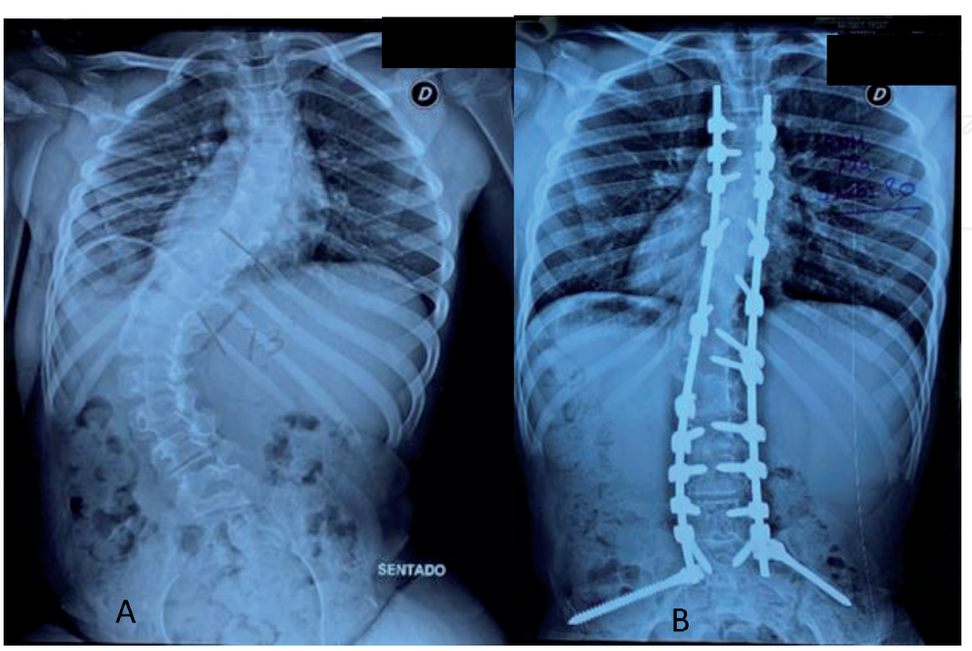

Figure 4.
Myelomeningocele patient radiographic image depicting scoliotic curve (A) and postoperative radiographic study depicting spinopelvic fusion (B).

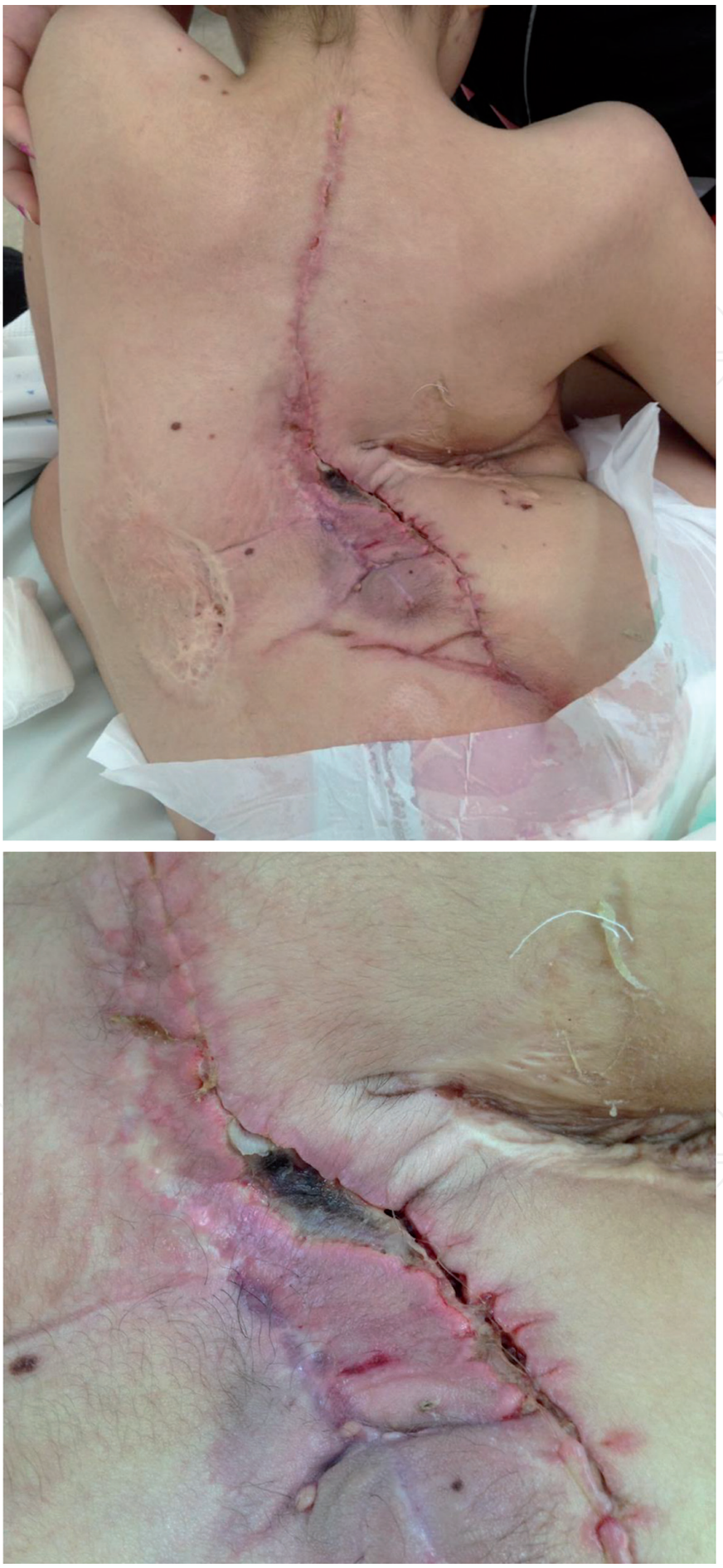

Figure 5.
Postoperative skin breakdown and infection in a myelomeningocele patient.

of correction, pressure sores, subsequent operations and even death (**Figure 5**) [21]. Some studies have suggested a higher rate of union when using a combined anterior and posterior approach [22].

Another spine problem spina bifida patients may present with is tethered cord syndrome. This occurs when the spinal cord is stretched because it remains attached distally, usually to scar tissue from prior surgical procedures. Most patients have some degree of cord tethering but only 30% manifest clinically. Patients who have symptoms present with progressive scoliosis, new gait abnormalities or changes, weakness, spasticity or back pain [23]. Neurosurgeons are the specialists who treat this problem surgically by untethering the cord.

5. Hip

Thirty percent of the spina bifida patients present with hip dislocations either at birth or during their childhood (**Figure 6**) [24]. The number can go up to 50% if we include hip subluxations. Dislocation occurs more commonly when the spinal cord defect is at the L3 level and the patient has a muscle imbalance with unopposed hip flexion and adduction. The ability of a patient to walk does not seem to be affected by dislocation of the hips and surgical relocation does not necessarily translate in a functional improvement [25]. Additionally, this problem does not seem to cause pain to the patients. For all these reasons many orthopedic surgeons advocate against putting the patients through complex osseous and soft tissue procedures and surgical intervention can even be considered controversial in such scenario where a benefit will not necessarily be obtained and such interventions are not exempt from surgical complications [26, 27].

6. Knee deformities

The most common knee problems spina bifida patients present with are knee flexion contracture and knee extension contracture [13]. Less commonly valgus deformity and instability [27]. There are many causes for those deformities such as muscle imbalance, fibrosis of the surrounding tissues and eventually a fracture malunion. A flexion contracture can usually be present at birth, different form

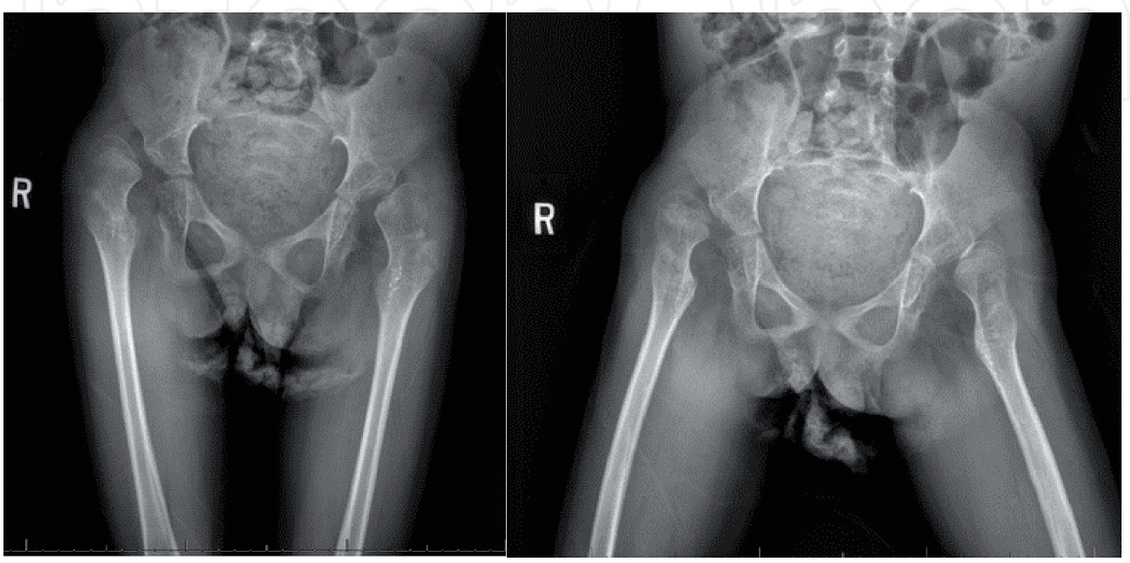

Figure 6.
Bilateral hip dislocation and osteopenia in a 14-years-old patient with spina bifida.

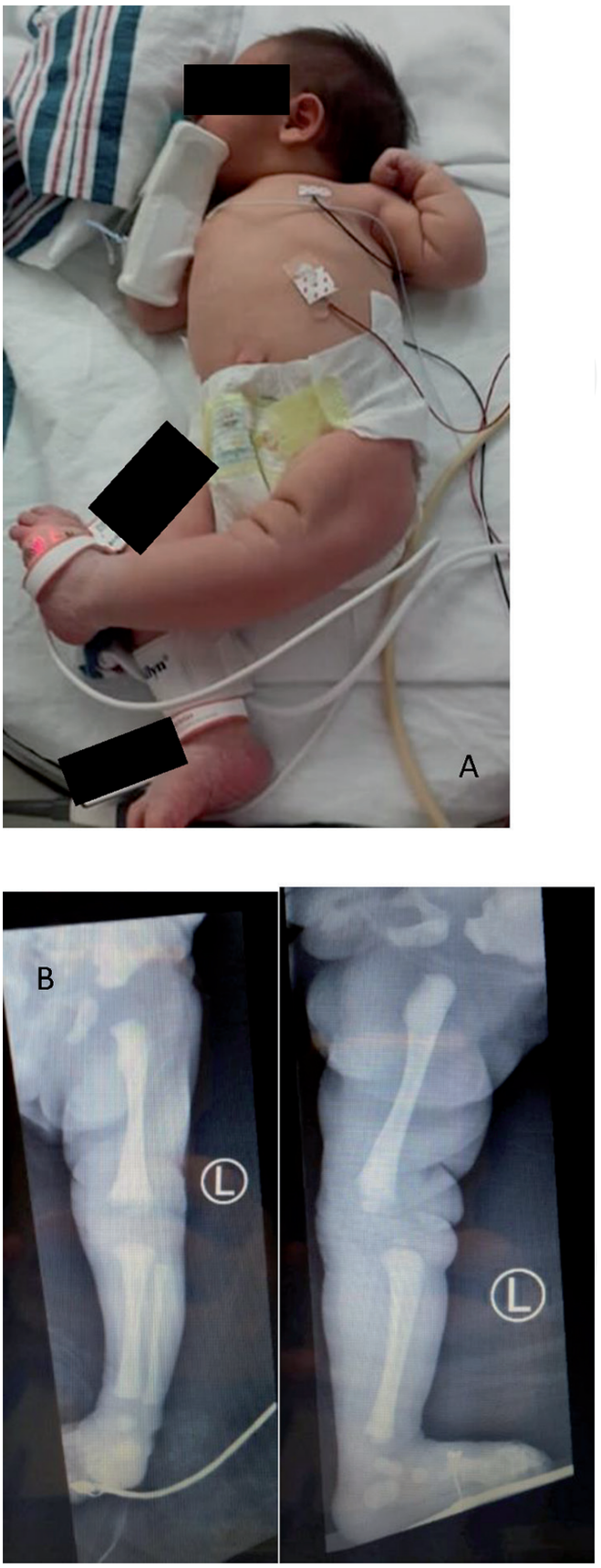

Figure 7.
Newborn with congenital knee dislocation in extension (A) and front and lateral radiographic image depicting the knee extension deformity (B).

the flexed knee found in healthy newborns, in myelomeningocele patients this deformity is fixed and more difficult to treat. The higher the level of the spinal cord defect the more severe is the knee contracture [28]. Patient positioning and muscle

imbalance are thought to be involved in the genesis of this deformity. If the patient is non-ambulatory the fixed knee flexion contracture does not cause any functional impairment, but in ambulatory patients it should be addressed. Surgical treatment is indicated when the flexion contracture is >20 degrees [29]. Treatment usually involves the releasing of the surrounding soft tissues such as hamstrings, gastrocnemius and posterior capsule. In more severe cases and usually in older patients an extension osteotomy may be indicated as well [30].

Knee extension is also usually present at birth, usually bilateral and much less common than the flexion contracture (**Figure 7**). The treating orthopedic surgeon should be aware of other associated deformities such as ipsilateral hip dislocation, external hip contracture and equinovarus foot [31]. If the patient presents with

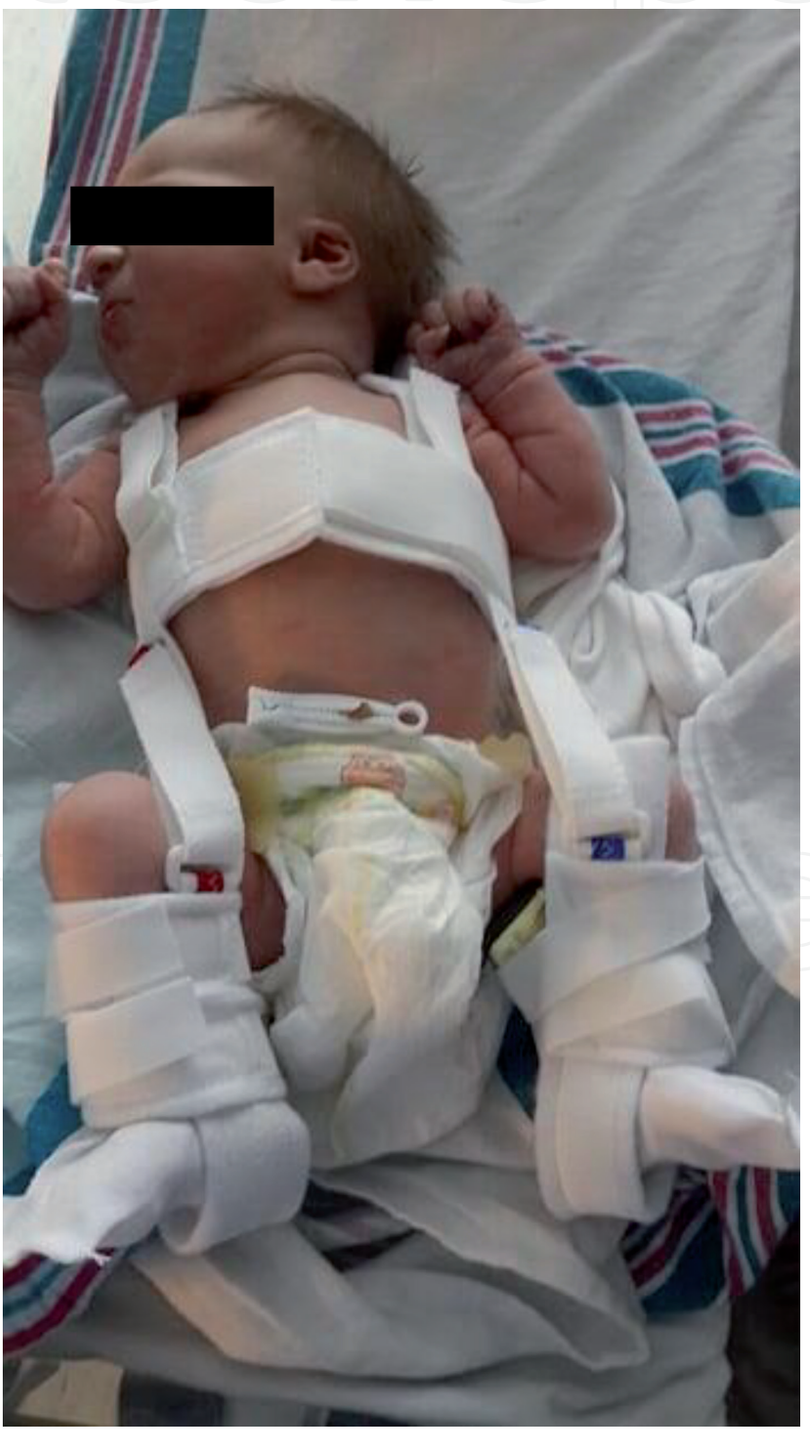

Figure 8.
Newborn wearing a Pavlik harness, the harness requires the knee to be bendable in order to fit appropriately.

Figure 9.
Extension deformity being treated by serial casting aiming a achieving a 90 degrees knee flexion.

hip dislocation and knee extension deformity simultaneously at birth, the knee deformity should be addressed first, so that the newborn can afterwards, once the knee deformity has been corrected, wear a Pavlik harness to treat the hip dislocation (**Figure 8**) [32]. The treatment for the knee extension deformity consists in serial casting until a 90 degree flexion is achieved (**Figure 9**). The treating orthopedic surgeon should be aware of not utilizing much force to flex the knee since the distal femur can be bent and even fractured in extreme cases. Casting should be followed by physical therapy. In resistant cases where casting is not successful surgical intervention is indicated. The surgical procedure usually consists of V-Y quadriceps lengthening and anterior capsulotomy [33].

7. Foot deformities

Foot and ankle deformity are very prevalent in spina bifida patients, with an incidence ranging from 60 to 90%. They can be present at birth or developed later on in life in close relationship with the spinal defect level [34]. In addition to the muscle imbalance and deformity the patients present with insensate feet which places a risk for skin breakdown and infections. The most common foot deformities are calcaneus, equinus, Varus, valgus, clubfeet and vertical talus and they can present as a single deformity or in combination [35]. Treatment of foot and ankle deformities is aimed at achieving a braceable plantigrade foot. In general treatment may start with casting or bracing and potentially a soft tissue surgical intervention to avoid fixed bone deformities. Once those are present osteotomies are needed to correct the foot. The patient needs to be examined regularly by a specialized pediatric orthopedist to detect tightness and incipient deformities can be early addressed (**Figure 10**).

7.1 Clubfoot

Spina bifida patients present with a rigid clubfoot deformity that is in general resistant to casting. This type of deformity can occur in up to 30–50% of the patients and the frequency increases with higher levels of the spine defect [36]. Casting with the Ponseti technique should be attempted and even though most of the patients achieve correction by this method almost 70% will relapse [37].

Figure 10.
Patient with a bilateral cavovarus deformity being examined in clinic with the help of a podoscope.

Additionally, if serial casting is being implement is paramount to assess skin integrity at every cast change in these patients due to their insensate feet. After correction is achieved by casting the treatment is followed by an Achilles tendon tenotomy, usually open in these patients [38]. If a wider soft tissue release is needed later on due to a recurrence a radical posteromedial release is recommended. In this procedure the subtalar, talonavicular and calcaneocuboid joints are completely released. After surgery casting followed by ankle foot orthosis (AFO) is required to maintain the correction. If a recurrence is then again noted, which may occur in 20–50% of the patients, a talectomy is indicated to achieve a plantigrade braceable foot [39].

7.2 Equinus

This deformity is also associated with higher levels of spina bifida. If the deformity is flexible an AFO may be attempted to prevent further progressing to a rigid equinus (**Figure 11**). With increasing severity of the deformity an Achilles tendon excision is recommended and even a radical posterior release if a plantigrade foot is not achieved after the Achilles resection [40]. Once the foot is an acceptable position a K wire is used in the talocalcaneal joint to maintain the alignment while to foot remains in a cast for at least 6 weeks (**Figure 12**).

7.3 Cavovarus

Cavovarus foot deformity is more prevalent in patients with a sacral level spina bifida and it is present in up to 17% of the patients [41]. The deformity is the cause of foot muscle imbalance (**Figure 13**). The treatment is dependent on how flexible the hindfoot is. This must be assessed by the orthopedic surgeon with the Coleman

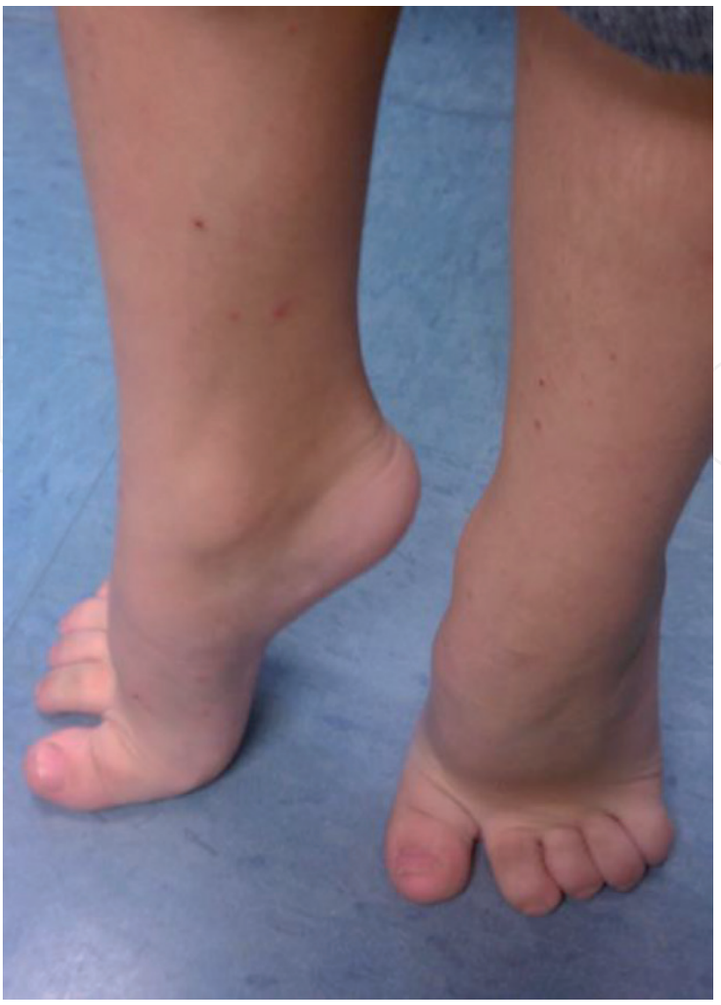

Figure 11.
Four-year-old patient with bilateral equinus.

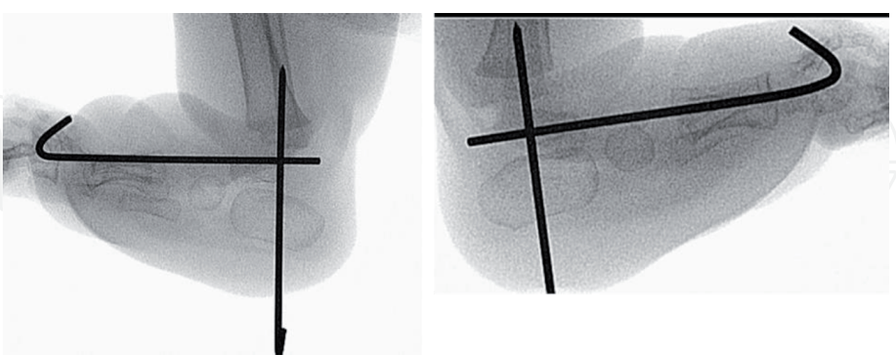

Figure 12.
K wires used after posterior release in a 12 months old patient with rigid bilateral clubfeet.

block test. If the hindfoot is flexible, only the forefoot will need to be addressed surgically. Meanwhile on the case of a rigid hindfoot several osteotomies may be needed to achieve correction. The current recommendations with high percent of success are for a first metatarsal closing wedge, an opening plantar wedge osteotomy of the medial cuneiform, a closing wedge cuboid osteotomy and sliding calcaneus osteotomy [42, 43].

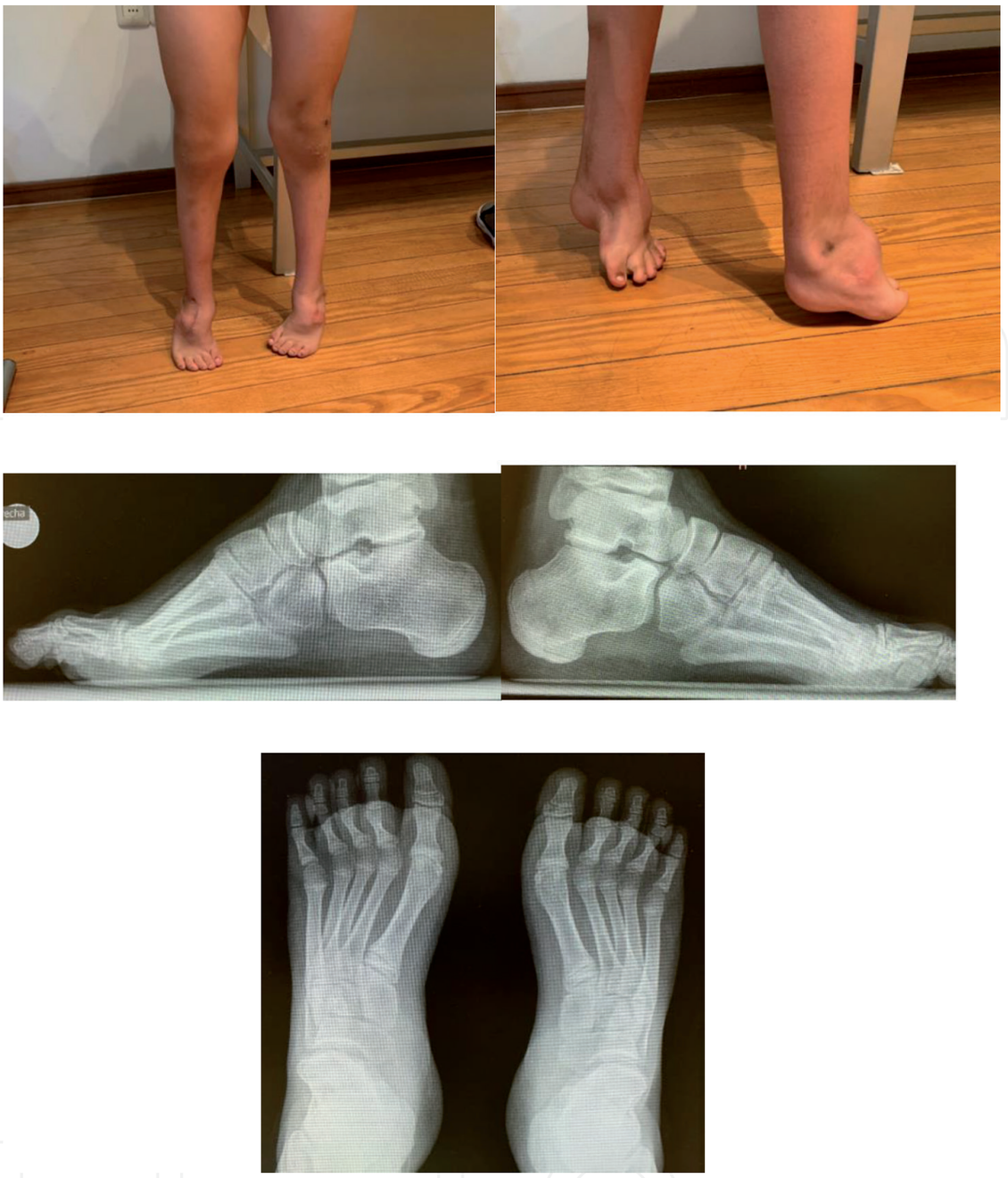

Figure 13.
Myelomeningocele patient with bilateral cavovarus feet and accompanying radiographic images depicting the high medial arch and the varus deformity.

8. Conclusions

Spina Bifida comprehends a complex subset of congenital malformation with a wide array of clinical presentation and truly diverse challenges to the patients affected by it. It is paramount that a team of multiple health care professionals from several areas of specialty work together to help improve the outcomes and life quality of these patients. The orthopedic surgeon is usually involved shortly after birth and continues to follow spina bifida patients for long terms into adulthood.

Conflict of interest

The authors state no conflict of interest related to the writing of this chapter.

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