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Spina bifida occulta in high grade spondylolisthesis

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Abstract. – A 14-year-old boy presented with symptomatic high-grade dysplastic type spondylolisthesis, with a presence of spina bifida occulta, not diagnosed by plain radiographs, but confirmed on preoperative CT and MR. Circumferential fusion with partial reduction of L5/S1 was performed. Awareness of the coexistence of spondylolisthesis and spina bifida by an accurate preoperative planning is paramount to avoid iatrogenic damage to neural elements during surgery.

Key Words:
Spina bifida, High grade spondylolisthesis.

Case Report

Introduction

Spondylolisthesis is a common etiology of back pain in children and adolescents. Surgical treatment is frequently instituted to manage symptoms and restore functions in selected cases.

The most popular and practical classification system in terms of prognosis and therapy is that of Marchetti and Bartolozzi[1]. In this system, spondylolisthesis is divided into two major groups, developmental spondylolisthesis (DS) or acquired spondylolisthesis (AS). DS is further divided into two types: low dysplastic (LDDS) and high dysplastic (HDDS), depending on the severity of the bony dysplastic changes present on the lumbar and sacral vertebrae.

HDDS can remain asymptomatic for a long time and can progress to a more severe grade of listhesis and spondyloptosis[2]; its congenital nature is supported by the fact that it is associated in one-third of patient[3] with spina bifida occulta of L5, of the sacrum, or of both[4].

Spina bifida occulta (SBO) is caused by failure of fusion between posterior vertebral elements without affecting the spinal cord or meninges. It is usually observed at the fifth lumbar vertebra and/or upper one or lower two sacral vertebras.

This possible co-existence should be carefully examined prior to surgery; if this is not done, significant risk exists for inadvertent damage to the neurologic elements during the approach to the posterior spine[5].

Background

Patients with DS usually develop symptoms during the prepubertal growth spurt, in adolescence[6]. The anatomic incompetence of the facet joints allows the slipping to begin as well as the biomechanical weakness of the sacral end-plate facilitates the progressive listhesis. In cases with SBO, lack or hypoplasia of posterior elements may increase the stress on pars interarticularis and lead to acquired deformities such as isthmic spondylolisthesis[7].

The aim of this paper is to describe a clinical example of HDDS in an adolescent male with unknown SBO and to illustrate the expedients to avoid neurologic lesions during partial reduction and fusion of HDDS.

Materials and Methods

A 14-year-old adolescent male, a student without any history of strenuous sporting activity, presented with worsening low back pain and developing lumbosacral kyphoscoliosis. His past medical history regarding episodes of back pain had been negative since 5 months before. Clinical examination showed lumbosacral kyphosis and compensatory left thoracic lordo-scoliosis. A palpable step-off at the lumbosacral junction, vertical position of the pelvis and sagittal unbalance were visible. Moreover he presented stiffness with hamstring shortening, flexed-hip, knee walking and toe gait. He also complained increasing radicular pain to the right buttock and thigh. Spine flexion and moreover extension were limited, the listhetic scoliosis rigid; neurological evaluation of the lower extremities showed bilateral quadriceps motor weakness. No bowel and bladder deficiencies, no clones were experienced (Figure 1).

Patient was referred to us with a diagnosis of spondylolisthesis made on the basis of plain radi-

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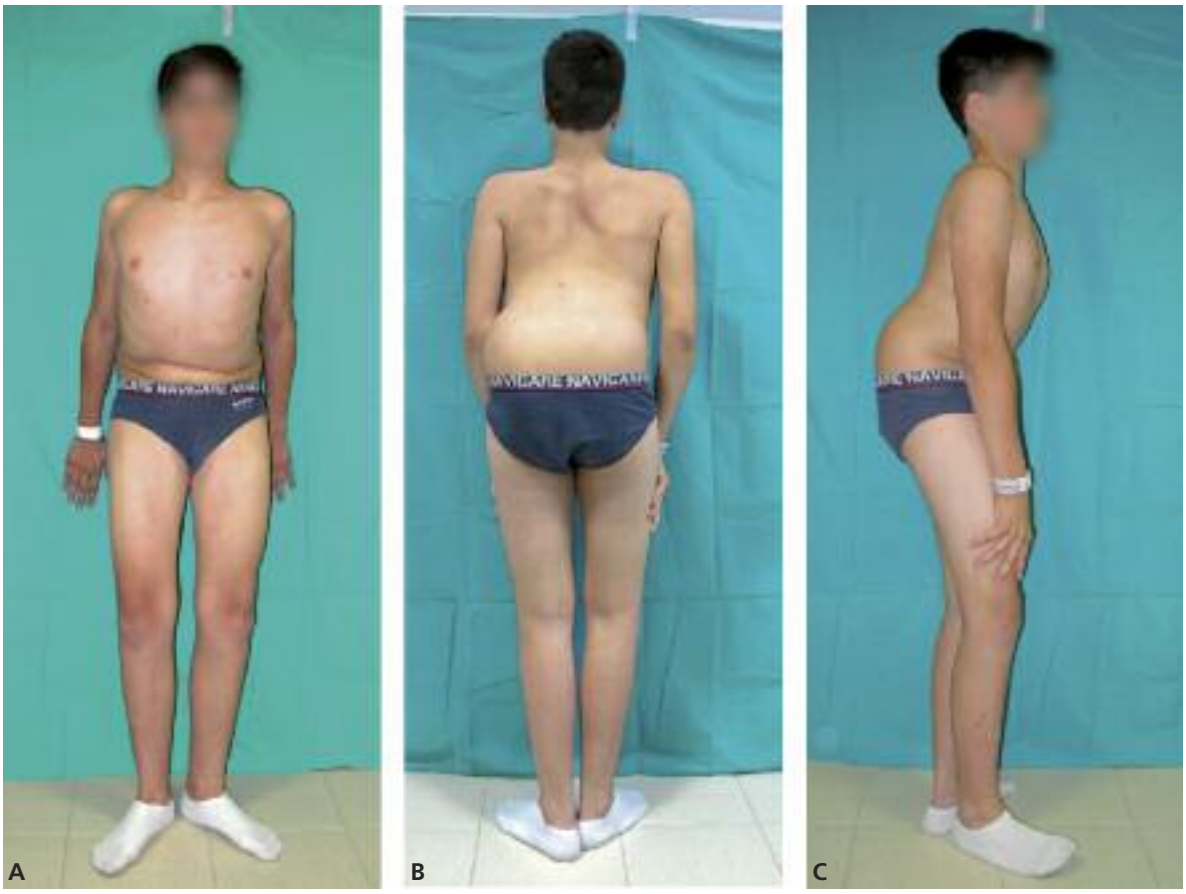

Figure 1. Clinical examination showed lumbosacral kyphosis and compensatory left thoracic lordo-scoliosis.

ographs, without any signs about other kind of dysplasia of the lumbar spine.

Plain radiographs showed Meyerding[8] grade III dysplastic spondylolisthesis at the L5-S1 level, lumbosacral insufficient lordosis (3°) with compensatory low grade left thoracic lordo-scoliosis (thoracic curve: 10°; lumbar curve: 13°) and unbalanced sagittal alignment with pelvis retroversion PT: 58, SS: 16, PI: 75 (Figure 2). It was also possible to detect a thoracic lordosis (-53°).

At X-Ray imaging S1 dysplasia was not adequately emphasized and further diagnostic studies with CT better highlighted the trapezoidal L5 body and its kyphotic tilt in relation to the sacrum, the significant bony defect in the posterior aspect, the presence of L5 spondylolysis (Figure 3).

MRI imaging allowed to confirm the diagnosis of SBO associated to HDDS, not well diagnosed on plain X-ray imaging (Figure 4). It was not clear if there was an anatomical plane between the fibrous tissue that substituted the posterior bony arch and the neurologic structures; this item is important to plan the correct surgery.

Conservative treatment was tried during three months, using a rigid brace and physiotherapy; it lead just to a little control of back pain, without any resolution of the neurological symptoms and signs.

The surgical solution was proposed and discussed with the patient and his family to explain the high risks of neurological damage.

L4-S1 fusion by posterior approach was selected and the treatment was performed on three steps:

1. Large decompression.
2. Partial reduction of the deformity and correction of sagittal balance.
3. Instrumented circumferential fusion by posterior approach.

Patient positioning in the operating room was a critical step in the procedure. With the patient lying prone, the hips were positioned in maximum possible flexion (60°-80°) allowing a partial reduction of the listhesis (Figure 5).

Through a midline skin incision, the paravertebral muscles were stripped from the spinous

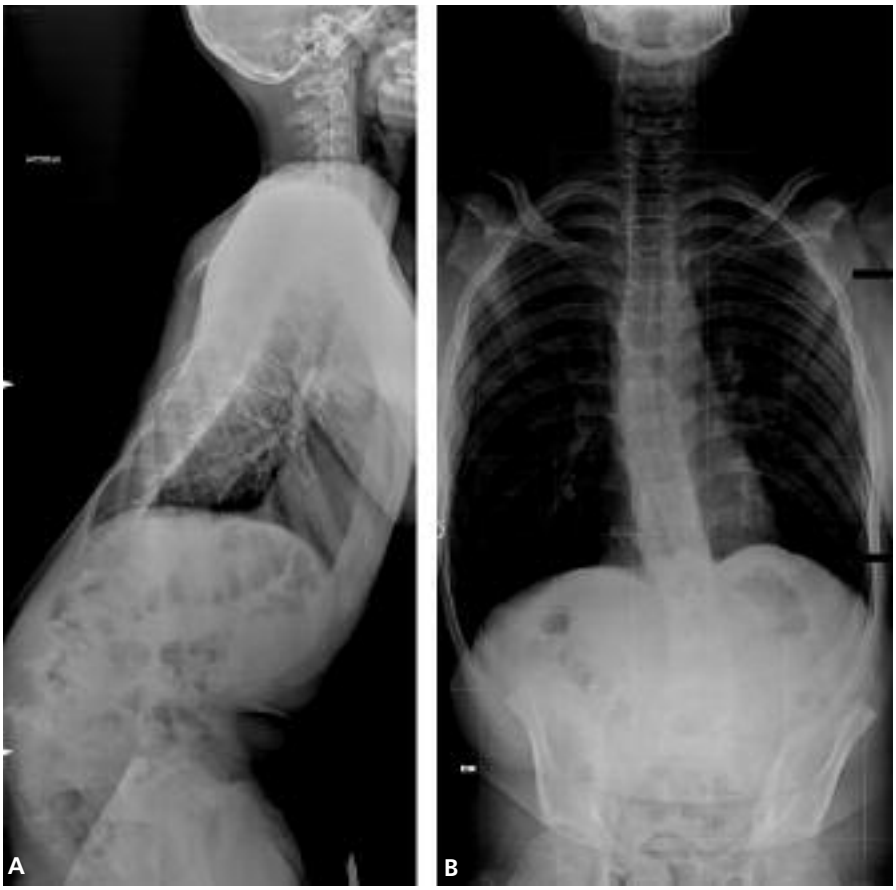

Figure 2. Plain radiographs, lateral ***(A)*** amd anteroposterior ***(B)***, showed Meyerding grade III dysplastic spondylolisthesis at L5-S1 level, lumbosacral kyphosis with compensatory left thoracic lordo-scoliosis and unbalanced sagittal alignment with pelvis retroversion.

processes. Pedicle screws were placed in L4, L5 and in the sacrum (bicortical in L5 and S1). The posterior elements of L5 and S1 were substituted with fibrous tissue that was completely removed to expose the L4 and L5 nerve roots until to their exiting from the foramen to ensure adequate visualization and avoid compressions during the reduction.

A lumbosacral discectomy was completed using a combination of disc space shavers, curettes

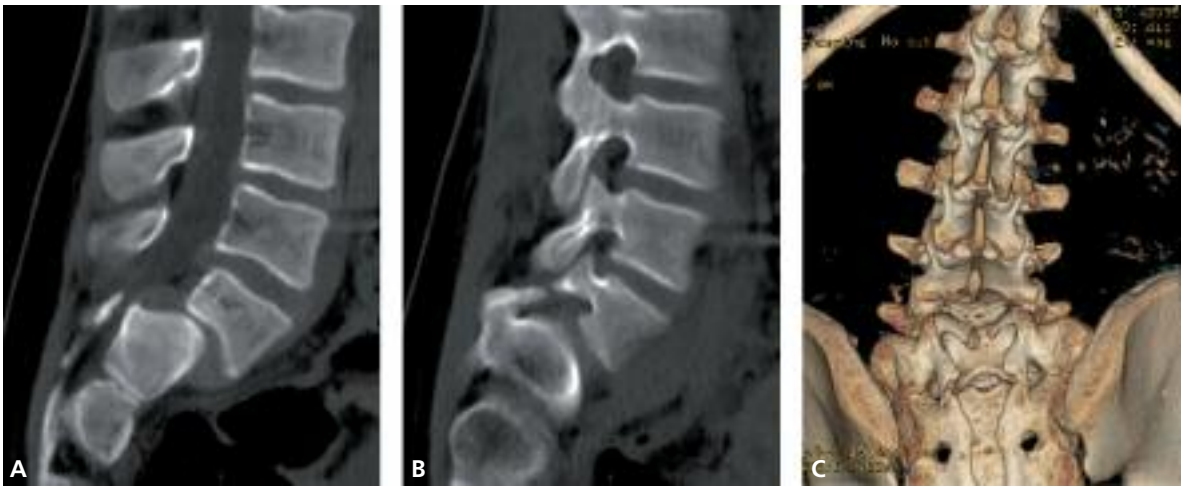

Figure 3. CT better highlighted the trapezoidal L5 body and its kyphotic tilt with respect to the sacrum, the significant bony defect in the posterior aspect, the presence of L5 spondylolysis.

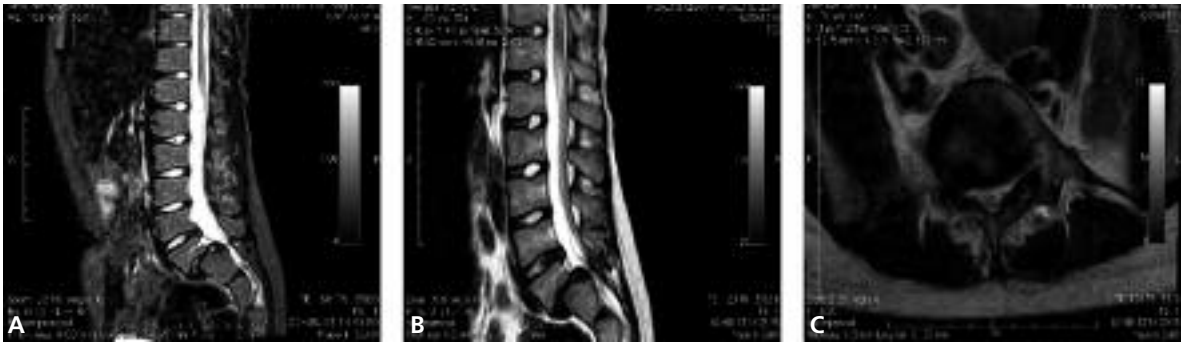

Figure 4. MRI imaging allowed to confirm the diagnosis of SBO associated to HDDS, not well diagnosed on plain X-ray imaging.

and rasps. The mobility obtained by the discectomy afforded additional reduction of the deformity.

Two rods were bent in the appropriate lordosis, tightened to the sacral screws and mounted to L4, L5 screw clamps without tightening.

At this moment, hips were extended to obtain the best possible reduction of the pelvic retroversion.

Gradual reduction of the listhesis was performed using the L4-L5 screws and rods system.

Interbody fusion was completed by autologous bone graft and one PEEK cage, while posterolateral fusion by decortication and grafting of the transverse processes of L4 and L5, and sacrum.

Results

Satisfactory reduction of the listhesis was obtained; frontal balance and sagittal balance (PI=65°, PT=40°, SS=25°) were partially restored (Figure 6).

Slight scoliosis in the thoraco-lumbar spine at frontal view (thoracic curve: 12°, lumbar curve: 13°) and hypokyphosis in the thoracic spine (-48°) at lateral view are still present. Lumbar lordosis improved to 13°. This compensatory posture is almost due to muscle spasm and will improve by proper physiotherapy.

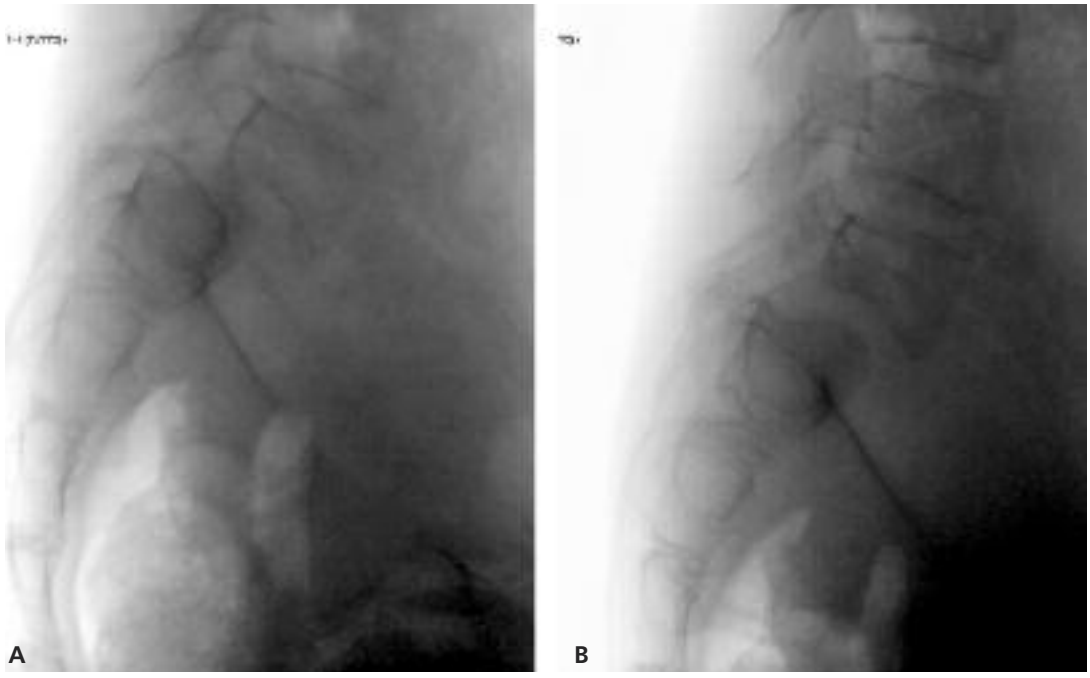

Figure 5. Preoperative dynamic X-ray. ***(A)*** During the flexion the spondylolisthesis worsens to grade III. ***(B)*** During the extension L5 aligns the sacral plate reducing the spondylolisthesis at grade I. This exam confirms the vertebral instability.

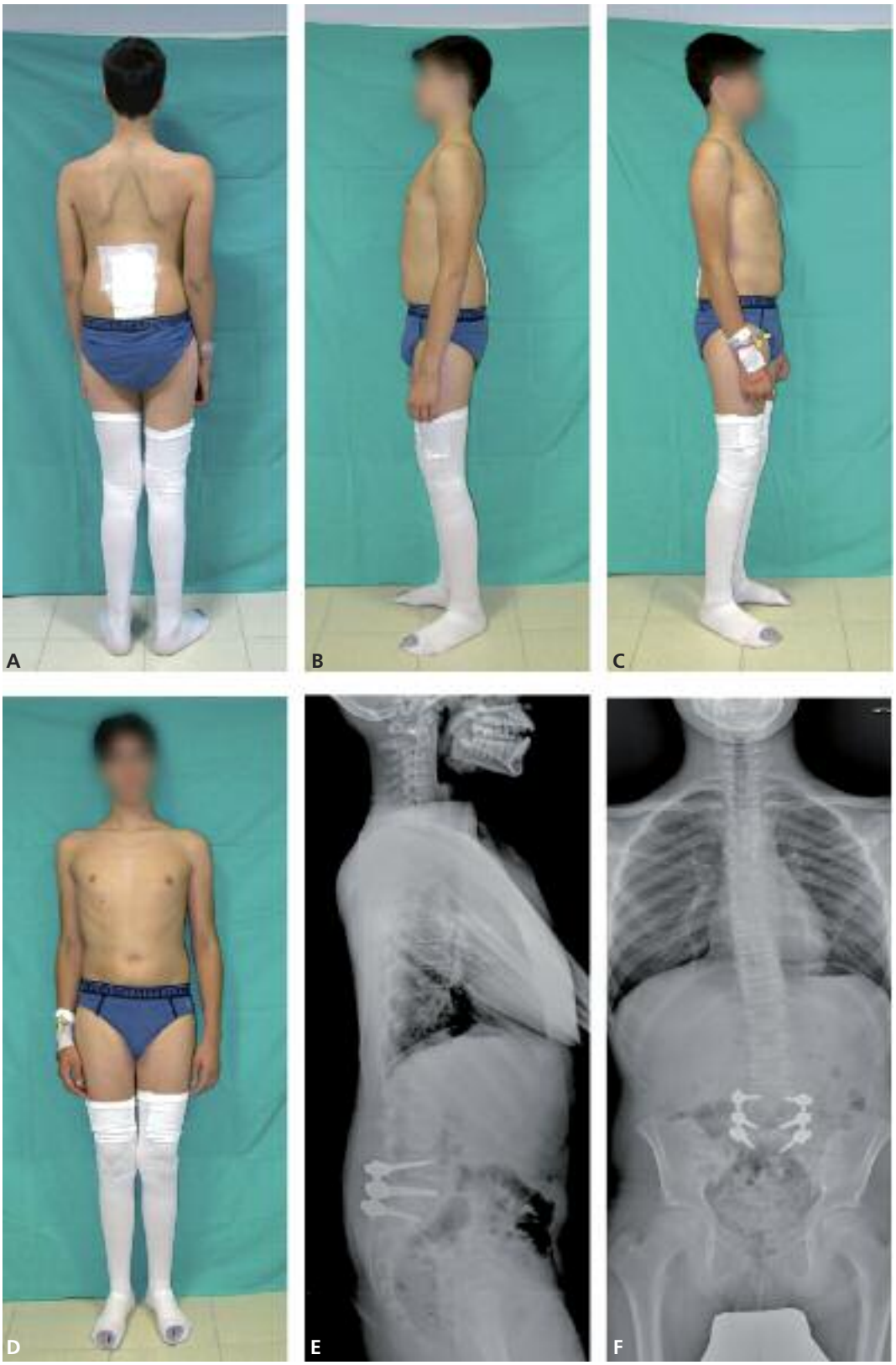

Figure 6. Postoperative pictures ***(A-D)*** and Postoperative lateral and anteroposterior X-ray ***(E-F)***, showing a satisfactory reduction of the listhesis. Frontal and sagittal balance were partially restored.

No major neither minor post operative complication occurred. Complete neurological deficits regression was obtained.

Discussion

Spondylolisthesis is a common etiology of back pain in children and adolescents. A variable amount of dysplasia, such as with spina bifida occulta, facet aplasia, or laminar aplasia, is common in spondylolisthesis. In particular, SBO is associated with spondylolysis of the lumbar spine in 11.8-35% of patients[9]. There have been reports of increased incidence of posterior spine defects in association with isthmic spondylolisthesis, although no etiological connection has been recognized. Blackburne and Velikas[10] and Hennrikus[11] found that the risk of a progressive vertebral slip was greater in patients with a midline lumbosacral defect. They proposed that this happened due to the lack of attachment of the multifidus muscles resulting from deficient spinous processes, which thereby decreased their stabilising effect. The presence of dysplastic or deficient posterior elements in the spina bifida defect may put increased loading on the pars[12-14] and lead to acquired deformities such as isthmic spondylolisthesis[5].

Dysplasia of the posterior arch in SBO is therefore to be considered as a risk factor of progression to high grade (> 50%) listhesis.

Treatment of spondylolisthesis of more than 50% in a growing child, as the case described, is usually operative[15]. Indications for surgery in high-grade dysplastic spondylolisthesis are mainly related to the following three factors: kyphosis, instability and progressive deterioration of listhesis with neurologic deficit[16,17].

As these procedures face major difficulties, many surgical techniques have been described, such as cast reduction and fusion, *in situ* fusion, laminectomy and *in situ* fusion, reduction and posterior instrumented fusion, reduction and posterior fusion combined with posterior interbody fusion[18], reduction and anteroposterior instrumented fusion with two different exposures, bilateral tubular minimally invasive approach for decompression, reduction and fixation[19], and vertebrectomy (Gaines procedure)[20].

The necessity of reduction and the degree of advisable reduction (partial or complete) is still a critical issue.

The current tendency is to reduce the listhesis only partially, especially in cases with preexisting neurologic deficits, to avoid worsening of the neurological preoperative situation.

Posterior stabilisation with decompression and 360° fusion showed the lowest incidence of pseudoarthrosis and is performed more frequently for reduction of high-grade spondylolisthesis.

The rate of neurological complications following reduction of high-grade spondylolisthesis is up to 25%[21]. These complications include nerve root injury (especially L5), cauda equina syndrome and injury to the superior hypogastric plexus (causing retrograde ejaculation in males and bladder problems and sexual dysfunction in females).

The importance of a coexistent dysplasia such as SBO lies in the greater danger of neurologic damage by surgical exposure and therefore it is recommend a detailed preoperative diagnostic approach of the disorder thorough an accurate clinical and imaging examination. The necessary imaging studies of the lumbar spine include plain radiographs, computed tomography and magnetic resonance imaging. Neurological evaluation is also mandatory. If this preoperative planning is not done, significant risk exists for inadvertent damage to the dura and roots during the approach to the posterior spine. SBO is indeed defined as the agenesia of the posterior vertebral arch with integrity of neurological structures. Surgical treatment is usually unnecessary, but when SBO is associated with other deformities to be treated, an inaccurate preoperative evaluation may be the cause of inadvertent neurological damage during surgery. Moreover, during HDDS treatment in cases of SBO co-existence, the fibrous tissue covering the dural sac in place of phisiological bone, must be removed to permit dural sac and roots control during reduction maneuvers and to free the structures themselves. This tissue can be very tight to the dura and sometimes to the roots, therefore preoperative imaging allows to visualize this connection and plan its excision. In our case we were lucky because the fibrous tissue that substitute the posterior arch was completely dived from the dura and a good plane of dissection was found to decompress the cauda; if the fibrous tissue appear attached to the neurological structures, a neurosurgical reconstruction of the dural sac should be planned and performed. This aspect should be accurately evaluated in the planning of the surgical intervention.

Conclusions

The unknown co-presence of high-grade spondylolisthesis and spina bifida occulta can be cause of intraoperative and postoperative complications. For these reasons it necessitates accurate preoperative planning, meticulous surgical technique and close postoperative monitoring of the patient. These will lead to a successful treatment outcome.

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Authors' declaration of personal interests

The authors declare no direct conflict of interests related to the specific topic discussed.

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