The Release Of Anchoring In Spinal Dysraphism

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Citation

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Abstract

Aim: In this study we briefly discuss the management of 7 different types of OSD leading to cord tethering. This study is focusing on the importance of releasing the fixing band(s) with a highlight on the value of intraoperative neurophysiological monitoring (IONM).

Materials and Methods: Retrospective study of 63 pediatric cases of OSD types. Documented full clinical and radiological assessment, followed by the surgical procedure aiming for release of the anchoring band(s). IONM was adopted in 41 cases. Second round of neurological assessment, with a follow up stages at three, six and twelve months.

Results: Retrospective study of 63 cases with OSD consisting of 30 males and 33 females, and with age ranging from one month till 9 years. In all cases microscopic exploration and identification of traction band(s) was performed. 41 cases had the chance of IONM guidance. No mortality, 3 cases with worsened neurological condition, 4 cases with fixed neuroglial condition after initial improvement, 30 cases with no added neurological deficit in comparison to preoperative condition and finally 26 cases with improved neurological condition.

Conclusion: Microscopic exploration and identification of traction band(s) considered mandatory in all cases of OSD. All the running data reinforce the use of IONM as a guideline of management of OSD.

INTRODUCTION

Spinal dysraphism refers to midline malformations of the back, from skin to vertebral column. It includes all lesions that result from incomplete formation of midline structures of the dorsum. It is classified as open form (operta) or closed form (occulta) (29). In occult spinal dysraphism (OSD) the dorsal skin is intact with no exposed neural tissue. The exact incidence of OSD in the general population is not entirely clear because many defects remain undiscovered and persist without being manifested. Yet 5–30% was a reported debatable rate and there is a significant female preponderance (3,5). Intraspinal anomalies that are commonly referred to as OSD include the split cord syndrome, fatty filum terminale, terminal syrinx, meningeocele manqué, dermal sinius tract, lipo-myelomeningocele, Neuroenteric cyst and myelocystocele (28).

Most manifestations of OSD are related to cord tethering i.e traction of conus medularis by anchoring bands. Prognosis of this syndrome is highly dependent on the degree of traction (19,25). Yamada, et al. have proposed that symptoms appear after hypoxic damage within the conus medullaris of patients with tethered cord syndrome (TCS) (30). Individuals with TCS may present with one or more combinations of myriad of symptoms and signs (4). Cutaneous lesions, including midline hairy tufts, hemangiomas, dermal pits/sinuses, hypertrichosis, subcutaneous lipomas, lumbosacral appendages, and nevi, are usually seen in 80 % of OSD patients (10, 20, 29). Neurological manifestations, which are due to disruption of motor and sensory pathways to the lower extremities, include gait disturbance, hyper-/hyporeflexia, muscular atrophy, spasticity, and sensory deficits (4, 8). Orthopedic abnormalities, including foot deformities, limb length discrepancies, gluteal asymmetry, scoliosis, and vertebral anomalies, such as bifid vertebrae, hemivertebra, and sacral agenesis, are found in 90 % of OSD cases (4, 29). Urologic symptoms range from incontinence, urgency, frequency, and recurrent UTIs to subtle changes observed on urodynamic studies (UDS). Early diagnosis of OSD in children is essential to attain better prognosis, so care providers must be

aware of the disease (4).

Current controversies with respect to the TCS include: 1) the early untethering of the spinal cord in asymptomatic patients; and 2) the proposed neurogenic hyperreflexic bladder resulting from a tethered spinal cord with a normally positioned conus medullaris. A majority of authors recommend early untethering procedure for the asymptomatic patient with TCS as early intervention might be an effective prophylaxis against progressive neurological and sphincteric decline, which may occur precipitously (6, 11, 16, 18, 24).

In this study we briefly discuss our experience and management results of different types of OSD in pediatric patients. This study is focusing on the value of releasing the fixing band(s) with a highlight on the value of intraoperative neurophysiological monitoring (IONM) as an essential aiding tool in the management of such cases.

PATIENTS AND METHODS

Patient population:

Sixty-three pediatric patients with different types of OSDs were retrospectively studied. The inclusion criteria for our patients included all patients under 18 years age that underwent appropriate surgical procedures at Ain Shams University Hospitals between January 2005 and January 2017 for treatment of different types of OSDs.

Clinical Assessment:

All patients underwent routine inspection for neurocutaneous stigmata and/or skeletal deformity of spine or lower limbs. Thorough preoperative neurological evaluation was performed for all our patients including full motor, sensory and reflexes in addition to gait assessment in patients that can walk. The presenting symptoms and neurological deficits were all recorded in a spreadsheet. Pediatric consultation was also done for all patients to search for any additional congenital anomalies.

Radiological Assessment:

Magnetic resonance imaging (MRI) involving the dorsolumbar region was performed for all our patients. MRI studies were essential for diagnosing and specifying the OSD type, for detecting the exact level of the conus medullaris, for revealing the nature of any subcutaneous swelling and also for predicting the anchoring factors that tether the conus medullaris. Computerized tomographic (CT) scans were done for selected cases with suspected split cord malformations or those with bony defects and malformations.

OSD Type:

Seven differetnt OSD pathoplogical types were represented and received treatment in our case series (Table 1).

Table 1

OSD types presented in our cases.

	OSD type	Number	Percentage
1	**Split cord**	15	23.8%
2	**Fatty filum terminal**	10	15.9%
3	**Syrinx**	12	19%
4	**Meningeocele manque**	3	4.8%
5	**Dermal sinus/Dermoid**	10	15.9%
6	**Lipomyelomeningeocele**	10	15.9%
7	**Neuroenteric cyst**	3	4.8%

Surgical Technique:

Sectioning of the Filum terminale, cutting the arachnoid and fibrous bands, releasing the spinal cord and correction of the associated malformation was the summary of the surgical technique that was adopted in all patients.

Patients were positioned prone under general anesthesia with supporting rolls on each side. Usually a midline back incision was used according to the nature and the level of the pathology. Dealing with the main pathology was performed first. In cases with lipomyelomeningeocele and filum lipoma the lipomatous masses were debulked as much as possible trying not to jeopardize any neurological tissues. Ultrasonic aspirator was used in some of our cases with lipomyelomeningeocele. Microscopic excision of dermoid tumors, dermal sinus tracts and neuroenteric cysts was also performed in cases having these pathologies. Removal of any bony spurs or fibrous septa was essential in the surgical management of cases with split cord malformations. With microscopic guidance, a midline dorsal durotomy was performed and dural edges were held up with stay sutures. Following the dural opening, filum terminale, arachnoid bands, and rootlets should be first observed. Introduction of microinstruments into the procedures should be used once the dura was opened. The most important issue at this surgical step was to differentiate the neural elements from any other extraneural structures. The filum terminale can be identified under the microscope by being a fibrovascular

cord like structure containing a large vessel, which becomes smaller across its course in the lumbar subdural space. After the surgical field becomes anatomically clear under the microscope, any suspected non-neural anchoring factor is coagulated and resected including thick filum, local spinal cord attaching bands, bony or fibrous septa, neoplastic/inflammatory adhesions and fibrous sinus tract. Following the detethering prodedure, the dura is tightly closed posteriorly with or without placement of a patch graft, whereas anterior dural defects were left open.

Intraoperative Neuromonitoring (IONM):

Forty-one patients of those enrolled in this study had the chance of being monitored intraoperatively. An ISIS neurophysiological monitoring system was used in our settings (Inomed, Germany). Preopertative bilateral insertion of intramuscular needles into: tibialis anterior muscles, gastrocnemius and anal sphincter is performed. After priming of the system, free running EMGs (frEMGs) were used for mapping to evaluate muscular firing during dissection.

Outcome assessment:

Records for surgically related complications, either early or late, were collected. All patients were subjected to postoperative full neurological assessment immediately postoperative, and at 3, 6 and 12 months after surgery during frequent visits. Radiological examinations were done in selected patients that required radiological follow up through out the follow up period.

RESULTS:

Patients demographics:

Sixty-three pediatric patients, that underwent surgical procedures at Ain Shams University Hospitals between January 2005 and January 2017 for treatment of different types of occult spinal dysraphism, were included in our retrospective study. The mean age of the patients at the time of operation was 50±26 months with a range from one up to 84 months. Thirty-three patients (52.4%) of those enrolled in the study were females and 30 (47.6%) were males (Table 2).

Table 2

Age and sex distribution for patients included in the study

Patients demographics	
Age	50±26 ms (1-84)
Sex	M n30 (47.6%) F n33 (52.4%)

Clinical Presentations:

Preoperative clinical assessments for our patients revealed that the main presenting symptom for our patients was either back or limb pain. Pain was evident in 69.8 % of our children. Motor and sensory deficits were manifested in 60.3% and 47.6% respectively. Sphincteric abnormalities including urinary and stool incontinence were found in 31.7% of patients. Twenty-two (34.9%) patients had different forms of cutaneous stigmata in association with other clinical manifestations. Nine patients had skeletal deformities in the form of dorsolumbar scoliosis (n=5), talipus equines (n=2) and both lower limbs length discrepancies (n=2) (Table 3).

Table 3

Clinical presentations for OSD patients in our study

Clinical Presentations	*Patient number*	*Percentage*
Axial Pain	*44*	*69.8%*
Motor deficit	*38*	*60.3%*
Sensory deficit	*30*	*47.6%*
Sphincteric disturbance	*20*	*31.7%*
Neurocutaneous stigmata	*22*	*34.9%*
Skeletal deformity	*9*	*14.3%*

OSD Type:

Seven different pathological OSD subtypes were represented in our case series. Frequencies were shown in table 1.

Clinical Outcome:

Of 44 patients that complained of back and leg pain 43 (97.7%) patients showed marked improvement during follow up period. Only one (2.3%) patient continued to complain of back pain throughout follow up visits. In the immediate postoperative period 3 (4.8%) patients showed newely developed neurological deficit whereas 60 (95.2%) patients

were stationary as preoperative status. At the end of follow up period 56 (88.9%) patients could regain full neurological function and on the contrary 7 (11.1%) patients had fixed deficits, four patients had worsened condition after intial improvent added to the three patients with early deterioration postoperative (Table 4). It worth mentioning here that only one (2.4%) patient of the 41 that were intraoperatively monitored was complicated by developing a new deficit early postoperative and continued to be a fixed tdeficit hroughout follow up.

Regarding the sphincteric control we had 20 patients that presented with urinary incontinence of which 5 patients had additional stool incontinence. In those patients with dual incontinence all the 5 patients showed only improvment in the stool incontinence with fixed urinary incontinence. In those with urinary incontinence only 2 (10%) patients reported transient improvement and worsening again in the last follow up.

Table 4

Neurological condition assessment throughout followup period

	Improved	Stationary	Worsened
Immediate post op.	--	60 (95.2%)	3 (4.8%)
3 month	26 (41.3%)	34(54%)	3 (4.8%)
6 months	60 (95.2%)	3 (4.8%)	--
12 months	56 (88.9%)	--	7 (11.1%)

Complications:

33 cases experienced postoperative CSF leak from the wound; 30 patients treated conservatively, 2 patients required repair once, only one case required repair 3 times.

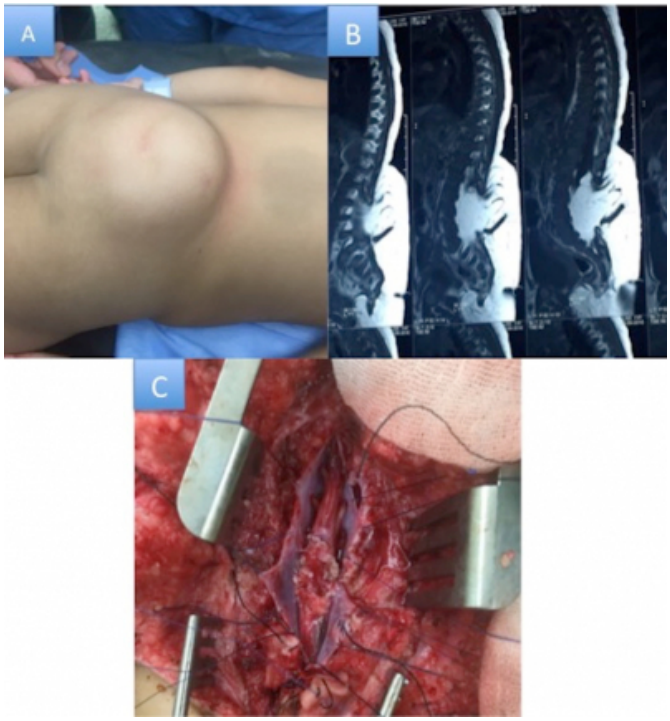
Case Presentation:

A 12 months old girl presented with a Doughy lump in the lower back that was noticed shortly after birth and increasing in size. She had Mild Rt LL weakness. IONM was set in this case. CUSA guided lipoma resection was done followed by filum terminale identification and resection. Direct posterior dural closure was performed.

Immediate post operative: No added deficit, lax subcutaneous collection with CSF leak that was managed by lumbar drain for 3 day. 3 months follow up; no neurological deficit. 6 months follow up: no neurological deficit. One year follow up: no neurological deficit (Figure 1).

Figure 1

A: Preoperative view for lumbar swelling in a lipomyelomeningeocele. B: MRI dorsolumbar spine showing the lipoma, dural defect and cord tethering. C: Intraoperative view after excision of lipoma.



DISCUSSION:

Although the incidence of spinal dysraphism has reduced (4) it is still common in the developing countries, due to lack of antenatal awareness and care (7,13). In this study we reviewed 63 pediatric patients that were operated upon for different types of OSD between 2005 and 2017. The mean age of our patients at the time of operation was 50±26 months with a slight female predominance. A wide range of congenital spinal anomalies can fall under the term OSD including split cord syndrome, fatty filum terminale, terminal syrinx, meningeocele manqué, dermal sinius tract, lipo-myelomeningocele, Neuroenteric cyst and myelocystocele (28). In our study 7 OSD types were presented with split cord malformation being most common (n=15), followed by terminal syrinx (n=12) and then comes fatty filum, dermal sinus tract with or woithout dermid and lipomyelomeningeocele each presented in 10 patients. A plenty of studies in literature have addressed the surgical management of different OSD types. There is still no consensus within the literature about the prevalence of different types of OSD (14, 24, 26).

Patients with different OSD types can present with a wide variety of signs and symptoms in combination with

cutaneous, orthopedic, spinal, sphincteric abnormalities, as well as pain (4). These manifestations are usually related to cord tethering by the filum terminale or anchoring bands (30). In our cases the most common cause for seeking medical advise was persistent back and leg pain (n=44). Neurological affection in the form of motor and sensory deficits was present in 38 and 30 patients respectively, while sphincteric disturbances were found to be present in 20 patients. Other important findings included cutaneous stigmata and skeletal deformity that were present in 22 and 9 patients respectively. In a study involving 49 pediatric patients with TCS resulting from different types of OSD conducted by Solmaz et al the patients symptoms included cutaneous lesions in 30 patients (61.2%), urinary disturbances in 10 patients (20.4%), leg or foot weakness, numbness and/or spasticity in 9 patients (18.4%), foot deformity (pes cavus and claw toes) in 5 patients (10.2%), scoliosis in 5 patients (10.2%), and back pain in 5 patients (10.2%) (23). In another retrospective study that reviewed spinal cord detethering in 61 children, 11 (18.0%) had motor or gait disturbance, 11 (18.0%) had sphincteric disturbance, eight (13.1%) had lower limb orthopaedic deformities, eight (13.1%) had scoliosis, six (9.8%) had back or leg pain and two (3.3%) complained of sensory disturbance (26).

Surgical management of tethered cord is still controversial among neurosurgeons. Patients with evident dysraphismic abnormality like lipomyelomeningeocele or those with clear neurological or sphincteric disturbances are considered straightforward candidate for untethering surgery. A lot of authors have recommended early surgery for patients who are neurologically intact for prophylactic purposes if tethering pathology is confirmed through a radiological investigation. There are many reports describing the surgery for tethered cord release in children (1, 12, 17, 26, 27). Most of these studies summarized the surgical technique as repair of associated lesion and cutting the filum terminale. But some surgical details are still missing in these reports. In our study, in addition to describing the surgical steps, our target aim is to highlight the microscopic detection and release of any suspected nonneural anchoring factor in the surgical field including thick filum, local spinal cord attaching bands, bony or fibrous septa, neoplastic/inflammatory adhesions and fibrous sinus tract. We could consider this step as a fundamental one in our approach to achieve sufficient untethering. We also introduced the intraoperative neuromonitoring as an essential aiding tool to differentiate between neural and non-neural tissues. Forty-one patients of those enrolled in this study had the chance of being monitored intraoperatively. In literature we could find also many studies that featured the use of intraoperative neuromonitoring during surgery for TCS (15, 21, 22).

Usually the overall clinical outcome for patients with TCS is dependent on the severity of the preoperative symptoms (9). The post-operative improvement in preoperative deficits in literature ranges between 40–80% (26). In a study conducted by Iqpal et al on 50 patients with TCS, 93% of patients showed improvement as regard leg and back pain. Motor weakness stabilized and improved in 57% and 42%, respectively. Untethering in patients with bladder or bowel incontinence improved in 46% of the patients (9). Thuy et al reported the results of their 5-year experience in the management of TCS in children. Their results were much worse than those reported in literature, where eight out of 30 children (26.7%) who had preoperative neurological, orthopaedic or sphincteric deficits were noted to improve post-operatively. Improvement in motor function or gait disturbance was documented at clinician follow up for four children (36.4% of those with symptoms), sphincteric disturbance for three (27.3%), scoliosis for one (12.5%) and back or leg pain for one child (16.7%) (26). Regarding our results we had 30 cases (47.6%) with no added neurological deficit. 26 (41%) cases had improved neurological condition. Three cases (4.8%) had worsened neurological condition. Four cases (63.5%) with fixed neurological condition after initial improvement. After 12 months of follow we could attain a final result of 89% neurological condition improvement and 11% worsening. Regarding the sphincteric abnormality we could report improvement in all the patients with stool incontinence whereas; only two patients of 20 having urinary incontinence showed transient improvement before worsening at one-year interval. All the patients that were monitored except one patient showed no added deficits. The delayed worsening in the clinical manifestations after initial improvement is usually attributed to retethering process.

The most common reported postoperative complications of untethering include cerebrospinal fluid (CSF) leakage, wound infection, meningitis, bladder dysfunction, and neurological deterioration (2, 4). An approximate range between 5–15% was reported for each surgical complication (9). In our experience although we had postoperative CSF leakage in 33 (52.4%) patients, 30 of them were transient and could be successfully managed conservatively by tight bandage, lying flat, dehydrating measures and CSF diversion by lumbar drain. Two patients required surgical repair and

duroplasty once. One patient in our series required 3 times repair to control the CSF leak. Fortunately no patients in this study suffered from overt CSF or wound infection that required further aggressive management. No procedure related mortality was reported in our patients.

One limitation of this study and other studies concerning OSD management is the heterogeneity of different pathological types falling under this term. This variability might confound the clinical results in different studies. Prospective case controlled studies are still deficient regarding the management of separate OSD types manifested with TCS. Such studies can provide us with high-level evidence for management of those cases; when to interfere, what to do and what to expect from surgery. From our experience we could recommend early surgical intervention for those patients presenting with cord tethering due to OSD. We herein also could conclude that using IONM renders the procedures more safe and effective.

CONCLUSION:

Occult spinal dysraphism includes a wide range of pathologies that usually manifest due to cord tethering. Early intervension is usually recommended for those patients manifesting with neurological or sphincteric disturbances. Microscopic exploration and identification of traction band(s) is considered mandatory in all cases of OSD. All the running data reinforce the use of IONM as a guideline of management of OSD, yet, no available research work strategy to state it as class A evident that IONM is standard in surgical management of OSD. Long term follow up for surgically treated patients is a must to early detect those who will manifest retethering of the cord.

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