Original Paper

Pediatric Neurosurgery

Pediatr Neurosurg 2008;44:360–370
DOI: 10.1159/000149901

Received: November 17, 2005
Accepted after revision: April 4, 2008
Published online: August 15, 2008

Correlation of Cutaneous Lesions with Clinical Radiological and Urodynamic Findings in the Prognosis of Underlying Spinal Dysraphism Disorders

Mehdi Sasani[a] Bahloul Asghari[c] Yalda Asghari[d] Ruya Afsharian[b]
Ali Fahir Ozer[a]

[a]Department of Neurosurgery, VKV American Hospital, and [b]Department of Obstetrics and Gynecology, Medamerican Medical Center, Istanbul, Turkey; Departments of [c]Neurosurgery and [d]Radiology, Tabriz Medical University, Tabriz, Iran

Key Words
Developmental abnormality · Spinal dysraphism · Cutaneous lesions · Human tail · Midline defective closure

Abstract
Spinal dysraphism is characterized by a lack of fusion of the vertebral arches that occurs in the absence of spinous processes with variable amounts of lamina. Here, we retrospectively present the importance of cutaneous lesions and their correlation with clinical presentation, radiological examination and urodynamic assessment. We retrospectively reviewed 612 (6.12%) cases with skin lesions from 10,000 consecutive live-born children seen at two institutions between January 1998 and March 2005. We divided all children into a control group and three groups based on clinical assessment, radiological examination and urodynamic evaluation results. Neurological deficits were identified in 113 (18.46%) children, while spinal dysraphism disorders, tethered cord syndrome and associated orthopedic malformations were found in 171 (27.94%), 119 (19.45%) and 28 (4.57%) patients, respectively. The incidence of tethered cord with lumbosacral dimple lesions over the sacrum (32/119, 26.65% of patients) was 3.5-fold higher than that of lesions found over the coccyx (9/119, 7.5% of patients). Uroneurological symptoms were found in 207/612 (33.82%) children. Urodynamic assessment revealed decreased bladder capacity in 10% of patients, detrusor hyperflexia during filling in 47% and a low-compliance detrusor in 71%. Discordance between ultrasonography and MRI was found in 16.58% of patients. Spinal cord untethering was performed for 109 patients. Nearly all children with resolution were at the end of their follow-up period (24 months). Retethering occurred in 21 (19.26%) patients, and a second untethering surgery was performed in 12 patients. When spina bifida was associated with lumbar skin lesions, there may have been an increased incidence of tethered cord and other spinal cord disorders. MRI scans are more reliable and give an exact diagnosis of tethered cord. Neurological and uroneurological instability are ultimately a clinical diagnosis, and there is controversy about their indications for surgery. However, the correlation between urodynamic assessment and cutaneous lesions with a tethered cord found by MRI examination allow for an early diagnosis and the possibility of prompt treatment.

Introduction

Spinal dysraphism, or spina bifida, is the congenital absence of a spinous process with variable amounts of lamina; it refers to a distinct group of developmental

KARGER

Fax +41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com

1016–2291/08/0445–0360$24.50/0

Accessible online at:
www.karger.com/pne

Mehdi Sasani
Neurosurgery Department, VKV American Hospital
Guzelbahce Sk. No:20
TR–34365 Nisantasi-Istanbul (Turkey)
Tel. +90 212 311 2760, Fax +90 212 311 2344, E-Mail sasanim@gmail.com

Table 1. Number and type of clinical presentations related to cutaneous lesions

Cutaneous lesions	Neurological deficits	Uroneurological problems	Orthopedic associated malformations			Syringomyelia
			pes cavus	congenital scoliosis	apparent limb shortening	
Lumbosacral dimple	35	83	3	1	–	2
Over the coccyx	7	24	–	–	–	–
Over the sacrum	28	59	3	1	–	2
Focal hypertrichosis	25	38	1	2	–	1
Strawberry nevus telangiectasia	20	19	6	1	1	–
Midline paraspinal subcutaneous mass	7	17	3	–	–	–
Lumbosacral lesions with dermal appendage tail	1	1	1	–	–	–
Multiple skin lesions	25	49	5	3	1	3
Total	113	207	19	7	2	7
Skin lesions (n = 612), %	18.46	33.82	3.10	1.14	0.32	1.14
Dysraphism abnormalities (n = 171), %	66.08	37.42	11.11	4.09	1.16	4.09
Neurological deficit (n = 113), %	100	56.63	16.81	6.19	1.76	6.19

anomalies characterized by a fusion failure of the midline structures derived from ectodermal and mesodermal tissues [1]. When spina bifida is associated with skin lesions, there may be a high incidence of dysraphism abnormalities such as tethered cords, lipomas and other spinal disorders [2, 3]. These disorders are not always obvious, as symptoms do not often manifest themselves until the infant becomes ambulatory. Symptoms leading to the diagnosis are upper motor neuron dysfunction, lower motor neuron dysfunction and their associated malformations [4]. There are 6 cutaneous manifestations of congenital neuroectodermal anomalies which are possible indicators of spinal dysraphism [5–7]; these include the presence of a dermal sinus (dimple), focal hypertrichosis, midline or paraspinal subcutaneous masses (low-lying lipomas) which cause gluteal cleft asymmetry, strawberry nevus or telangiectasia, cutaneous appendage or rudimentary tail, and atretic meningocele associated with abnormal gait and lower limb posture changes [2, 8–11]. About 50% of these lesions are associated with spinal dysraphism disorders and therefore are of great diagnostic value [12]. The other group of symptoms is caused by tethering of the spinal cord [4]. Congenital lumbosacral lipoma, diastematomyelia, fibrous adhesions and a shortened filum terminale cause conus entrapment in spinal dysraphism disorders [13]. Bladder dysfunction represents another group of symptoms. Early diagnosis with ultrasonography (USG), CT scan and/or MRI is mandatory for the examination [14, 15]. MRI has the important role of determining the presence of spinal dysraphism and its associated disorders because of its high specificity and resolution. Following diagnosis of these developmental abnormalities, appropriate surgical and medical treatment can be instituted [16, 17].

In this study, we present a series of patient outcomes and investigate the correlation of cutaneous lesions with USG and MRI findings in the determination of spinal dysraphism and underlying intraspinal dysraphism abnormalities. We also describe the results of our urodynamic assessments of infants with closed spinal dysraphism disorders.

Materials and Methods

We retrospectively reviewed medical records as well as operative, USG, MRI and urodynamic reports. To limit the chance of error, the data from all consecutive live neonates were also compared with databases from obstetrics-gynecology. The records from 10,000 live-born neonates delivered at two institutions between January 1998 and March 2005 were reviewed. All neonates were studied via clinical assessment of cutaneous lesions, neurological examination of upper and lower motor dysfunction, uroneurological symptoms, and associated orthopedic malformations by a multidisciplinary evaluation committee. This committee was formed by a neurosurgeon, pediatric neurologist, urologist and orthopedist. Databases used in this study were approved by an independent ethical review board at two institutions. Neurological examinations were carefully performed in all neonates by two pediatric neurologists at separate institutions, and then they reviewed the data together so that all the neurological examinations were standardized. Motor (muscle power), sensory (pin prick in the lower extremities and perineal area) and deep tendon

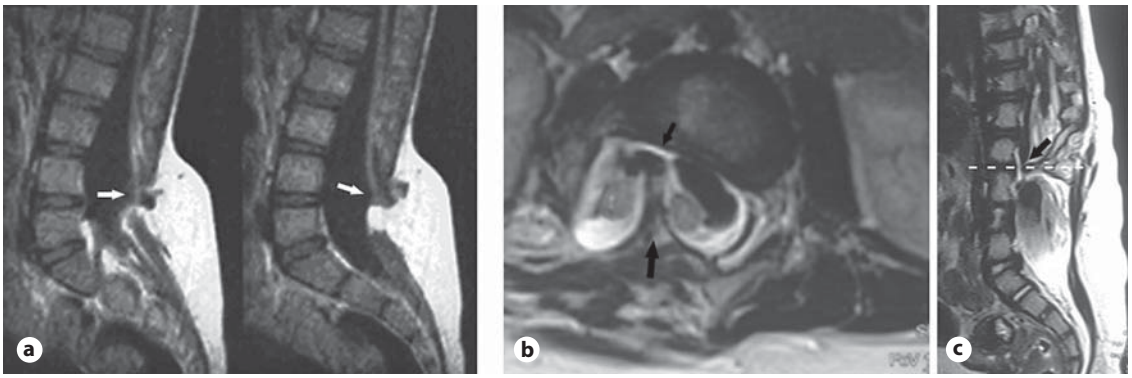

Fig. 1. a Sagittal MRI view; subcutaneous lipoma mass with tethered cord (white arrows). **b**, **c** Axial and sagittal MRI view of a diastematomyelia; black arrows show bone spur.

reflexes were included in the neurological assessment (table 1). A detailed urinary questionnaire was filled out for each child. Although the uroneurological examination was difficult, all parents were educated on the procedural details and were well informed. The clinical findings in this survey have been enumerated not only to catalogue their prevalence, but also to emphasize how dysraphism disorders affected their parents and their primary physicians. The questionnaire included indications regarding the phase of symptoms (such as diurnality more than 8 times), voiding difficulty and urinary retention. For ease, we initially divided all neonates into two major groups before clinical assessment based on the presence of skin lesions, which was further characterized by the presence of neurological deficits, urodynamic assessment and radiological examination (table 2). The first group (I-G) was neonates without lower-back cutaneous markers.

The records from group I-G were the medical records of neonates without lower back cutaneous markers that were judged to contain adequate information. Among 9,388 neonatal records, 760 neonates were considered for the control group ($I\text{-}G_c$).

The second group (II-G) consisted of neonates with cutaneous lesions at the midline of the lower back. This group was classified into the three following subgroups: one for the presence of neurological examination findings (II-GN), one with neurological deficits ($II\text{-}GN_a$) and the other without neurological deficits ($II\text{-}GN_b$). The other two groups of II-G were divided by spinal USG and/or MR scan findings (II-GR) and urodynamic assessment results (II-GU). The first subgroup of the radiological examination findings, group $II\text{-}GR_a$, consisted of neonates with cutaneous markers at the midline of the lower back which were associated with radiology findings of spinal dysraphism disorders. The second subgroup was neonates with cutaneous markers without spinal dysraphism disorders ($II\text{-}GR_b$). In radiology medical reports, we found that a number of neonates with cutaneous lesions were evaluated by USG and many of them were evaluated again by MRI (fig. 1). The first-line screening test was USG examination that was performed around the first birthday. Follow-up radiological examinations were performed with USG or MRI between 1 and 24 months of age for initial spinal disorders. If the USG was equivocal or abnormal, the neonates were re-evaluated by MRI scans. Urodynamic studies were performed in all infants with skin markers associated with spinal disorders in the $II\text{-}GR_a$ subgroup and in many infants of the $II\text{-}GR_b$ subgroup that had skin markers without spinal disorders. The first urodynamic assessment was performed between 3 and 24 months of age with spinal dysraphism disorders determined by radiological investigations. Depending on the age of the patient (infants aged 3–24 months), urodynamic assessment was performed with the child sitting or recumbent, under sedation. The urodynamic studies were usually assessed by a small electrode that was placed on the infant's hip, while the other end of the wire connects to a computer and also to two electrodes that were placed near the opening of the rectum. A thin catheter was inserted through the urethra and was used to fill the bladder with water and measure how much fluid the bladder held and how well it emptied. Another tube with a balloon on the tip was placed just inside the infant's rectum to measure pressure. Documentation related to the urodynamic study was assessed to evaluate detrusor overactivity compliance,

Table 2. Ratio and classification of study groups (10,000 live-born neonates)

I-G: First major group
I-G: 9,388 neonates (93.88%) without back-midline skin markers
$I\text{-}G_c$: control group, 760 neonates
II-G: Second major group; 612 (6.12%) with lower back-midline skin markers
Neurological assessment II-GN
Group $II\text{-}GN_a$: with neurological deficit (n = 113, 1.13%)
Group $II\text{-}GN_b$: no neurological deficit (n = 499, 4.99%)
Radiological examination II-GR
Group $II\text{-}GR_a$:with spinal pathology (n = 171, 1.71%)
Group $II\text{-}GR_b$: without spinal pathology (n = 441, 4.41%)
Urodynamic assessment II-GU
Group $II\text{-}GU_a$ (n = 207, 33.82%)
Group $II\text{-}GU_{a1}$: with pathological findings (n = 53, 25.60%)
Group $II\text{-}GU_{a2}$: physiological result (n = 154, 74.40%)
Group $II\text{-}GU_b$: none performed (n = 405, 66.18%)
Urodynamic assessment was performed in 207 neonates.

Table 3. Incidence of neurological deficit in dysraphism disorders, tethered cords and radiological examination findings related to various cutaneous lesions

Cutaneous lesions	Cases	Ratio to all neonates (n = 10,000)	Ratio to neonates with skin lesions (n = 612)	Neurological deficit			Evaluation with both USG + MRI (n = 250)		Tethered cord (neurological evidence and radiological findings: positive)					Tethered cord (uroneurological/ neurological evidence positive, radiological findings normal, anatomical conus medullaris normal)	
				cases	ratio to all neonates (n = 10,000)	ratio to neonates with skin lesions (n = 612)	agreement between USG and MRI	discordance between USG and MRI	cases	thickened and fatty filum terminale	diastematomyelia	lipomyelo-meningocele	dermal sinus tract	neurological deficit	uroneurological deficit
Lumbosacral dimple	264	2.64%	43.13%	35	0.35%	5.2%	62	14	41 (34.45%)	21	3	1	16	–	1
Over thecoccyx	156	1.56%	25.49%	7	0.07%	1.14%	22	3	9 (7.5%)	7	–	–	2	–	–
Over thesacrum	108	1.08%	17.64%	28	0.28%	5.7%	40	11	32 (26.95%)	14	3	1	11	–	1
Focal hypertrichosis	204	2.04%	33.33%	25	0.25%	4.08%	55	5	21 (17.64%)	12	–	–	9	–	–
Strawberry nevus telangiectasia	62	0.61%	10.33%	19	0.19%	3.10%	16	–	20 (16.80%)	14	1	–	5	–	–
Midline paraspinal subcutaneous mass	32	0.32%	5.55%	7	0.07%	1.13%	10	6	15 (12.63%)	7	1	7	–	–	2
Lumbosacral lesions with dermal append-age (tail)	1	0.01%	0.16%	1	0.01%	0.16%	1	–	–	–	–	–	–	1	–
Multiple skin lesions	49	0.46%	7.5%	26	0.26%	4.24%	27	9	22 (18.48%)	12	6	2	2	–	–
Total	612	6.12%	100%	113	11.3%	18.5%	171 (27.94%)	34	119 (19.45%)	66	11	10	32	1	3

residual urine, bladder capacity and detrusor leak point pressure. The patients with severe neurological deficits associated with tethered cords, as determined by USG and MR scans, underwent surgery and urodynamic studies were also done before surgery.

Fifty-eight female and 51 male children with tethered cords who were between 6 and 19 months old (mean age 1.2 years) underwent surgical treatment. Surgical procedures were performed for the underlying dysraphism disorders for the untethering and release of the spinal cord by two neurosurgeons in the two institutions. Follow-up examinations were done 3 months after surgery and then every 3–24 months depending on the clinical assessment and the urodynamic study evaluations. In a review of the patient charts, we found that the patient assessments of the two participating institutions for the treatment of these neonates were the same. Both of them followed a standard approach for closed spinal dysraphism disorders.

Results

Control Group Findings

The records from 9,388 neonates without lower-back skin markers (group I-G) were reviewed, in which we found 760 neonates who were evaluated with USG or MRI examinations. These neonates had no skin markers, and neurological examinations were free of any neurological findings. The urodynamics evaluation was not performed for this group. We did not clarify why these neonates underwent radiological examinations, but they had no lower back skin lesions with normal neurological findings. We found 83/762 (10.8%) of neonates with a lamina ossification defect. All neonates were examined with USG and lumbar MRI. We found 83/762 (10.8%) of neonates with lamina closure defects. It was unexpected that all 762 neonates underwent both USG and MRI examinations. First, they were examined by USG and then they underwent MRI examinations. USG findings and MRI findings were in agreement, and all of the control group patients only had lamina disclosure defects without spinal cord disorders and skin markers.

Clinical Findings

We found lower-back cutaneous lesions in 612 (6.12%) neonates. The cutaneous lesions were dermal sinus dimples in 264 (43.13%) neonates. These lesions were located over the coccyx (156; 25.49%) and over the sacrum (108;

17.64%). In addition, hypertrichosis was present in 204 (33.33%), strawberry nevi or telangiectasia were present in 62 (10.33%), midline or paraspinal subcutaneous masses were present in 32 (5.55%), a cutaneous appendage named 'human tail' was present in 1 (0.16%), and multiple skin markers were present in 49 (7.5%) neonates (table 3, fig. 2). Only 1 case had a cutaneous appendage (human tail) about 7.2 cm long in the lower lumbar area. Pathological examination of the tail showed subcutaneous fat and areolar tissue with some nerve fibers covered by skin.

A hundred and thirteen neonates (18.46%) had neurological deficits. These deficits presented as upper motor neuron dysfunction in 63 (55.75%) and lower motor neuron dysfunction in 50 (44.24%). The neuropathological signs included lower-extremity weakness in 41 (36.28%), muscle tone abnormalities in 28 (24.77%) and sensory disturbances in 19 (16.81%). Orthopedic associated malformations included pes cavus in 19 (16.81%), congenital scoliosis in 6 (5.30%) and apparent limb shortening in 2 (1.76%).

Uroneurological Symptoms and Urodynamic Evaluations

The urinary questionnaire was filled out for 612 patients with skin lesions. We found that 207 of 612 patients had uroneurological symptoms. All patients with tethered cords had significant uroneurological symptoms. The most common symptoms were voiding difficulty in 63% of patients, filling phase symptoms in 69% of patients and urinary retention in 4% of patients. Urodynamic assessments were also performed for 207 neonates. The normal volume of residuals in children participating in the study (control group with an age range of 3–24 months) was less than 30 ml. For comparison, the physiological volume of residuals is normally below 10% of total bladder volume. The average residual volume was 125 ml in 81 patients (39%). Decreased bladder capacity was noted in 20 (10%) patients. Detrusor hyperflexia during filling was found in 97 (47%) patients, and low detrusor compliance was found in 145 (71%).

Radiological Evaluation

Neonates with lower-back midline lesions (group II-G; n = 612) were investigated by radiological examination, and 407 (66.50%) of them had normal USG findings. Neonates that were suspected of or detected with spinal dysraphism disorders (205, 33.50%) were examined with USG and then were evaluated with MRI. Spinal dysraphism, which is associated with dysraphism disorders, was found by both USG and MRI in 171 (27.94%) of the 612 cases with skin markers. Spine MRIs for 34 of 205 neonates were done with questionable USG findings. The subsequent MRI demonstrated no spinal disorder; therefore, 34 cases (16.58%) of the 205 neonates in this group had disagreement between USG and MRI findings (table 3). Consequently, 441 (72.06%) neonates had no dysraphism disorders, and 171 (27.94%) neonates had different spinal dysraphism disorders based on the MRI examination findings.

Spinal dysraphism was associated with a thickened fatty filum terminale in 101 (59.06%), diastematomyelia in 11 (6.43%), lipoma and lipomyelomeningocele in 10 (5.8%) and dermal sinus tract in 49 (28.7%) neonates.

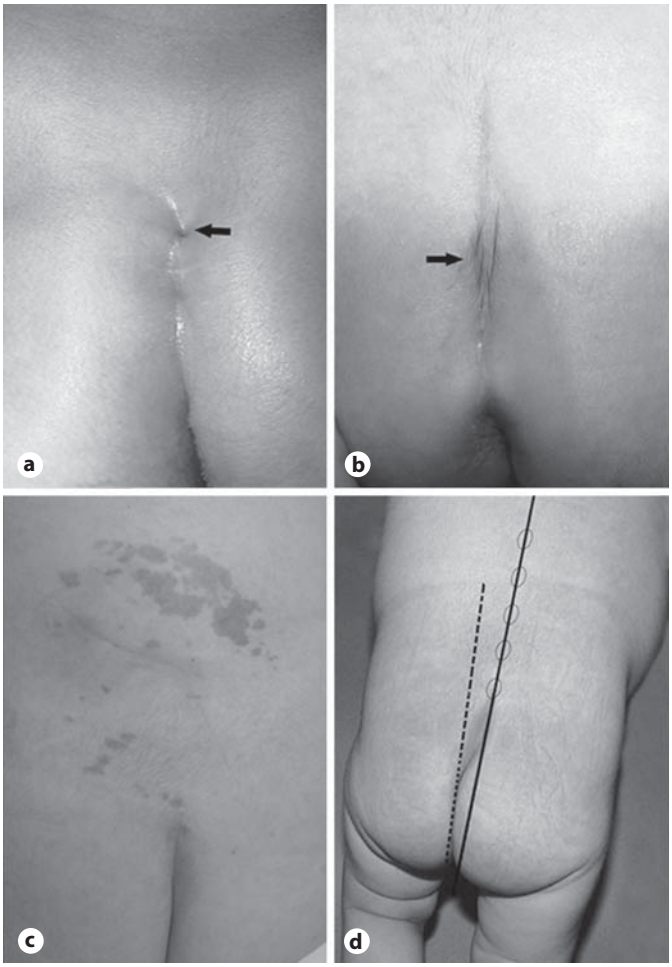

Fig. 2. Lower-back cutaneous markers. **a** Skin dimple. **b** Hypertrichosis. **c** Capillary hemangioma and hyperpigmentation. **d** Subcutaneous lipoma mass causing midline gluteal line deviation.

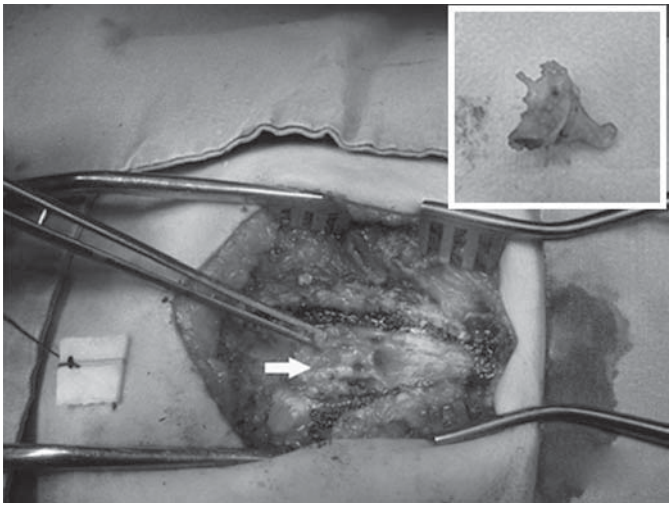

Fig. 3. Resection of bone spur in a diastematomyelia patient; place of bone spur viewed during operation (white arrow).

Tethered Cord Syndrome

Among the 171 cases with dysraphism disorders that were determined by radiological examinations, 119 neonates were found to have a tethered cord. Of these 119 neonates, 6 were asymptomatic and 113 presented with neurological deficits. Therefore, 1.19% of live-born neonates and 19.5% of neonates with cutaneous lesions had a tethered cord. There were 41 neonates with tethered cords and lumbosacral dimples; of these, a thickened fatty filum terminale was present in 21/41 patients, diastematomyelia in 3/41, lipomyelomeningocele in 1/41 and dermal sinus tract in 16/41. A tethered cord with lumbosacral dimple lesions was present in 41 (34.5%) patients: 9 neonates had a lumbosacral dimple over the coccyx, and 32 neonates had it over the sacrum. Consequently, the incidence of tethered cord with lumbosacral dimple lesions over the sacrum was 3.5-fold more prevalent than lesions over the coccyx. Among the remaining 78 neonates with skin lesions associated with tethered cords, 21 (17.60%) also had focal hypertrichosis, 20 (16.8%) had strawberry nevi and telangiectasia, 15 (12.60%) had midline paraspinal subcutaneous masses, and 22 (18.5%) had multiple skin lesions (table 3).

Clinical Evidence of a Tethered Cord with Normal Radiological Findings

Four children presented with the neurological or uroneurological findings of a tethered cord syndrome; however, there was disagreement between the clinical examination and radiological evidence (table 3). In all 4 children, the conus medullaris had a normal anatomical ending. One patient had clubfeet and paraparesis with a cutaneous appendage (human tail). Three patients had uroneurological findings with 1 of 3 patients having a dimple over the sacrum, while 2 of 3 patients had a lipoma and a normal anatomical ending. For treatment, 1 patient underwent surgery. Three patients did not have surgery and were followed up with conservative treatment. One of 3 patients for whom conservative treatment was applied gradually worsened. Two of the 3 nonsurgical patients and 1 patient who underwent surgery showed improvement.

Surgical Findings

All 119 neonates with low-lying conus and tethered cords had syndrome symptoms, and with the exception of 10 patients, they underwent surgical treatment. These 10 patients had neurological deficits associated with tethered cords, and surgical treatment was required, but their parents did not agree to surgery. The remaining 52/171 neonates with spinal disorders without neurological deficits and tethered cord syndromes were managed without surgery. The varying spinal dysraphism disorders which caused tethered cords associated with skin lesions were operated on via different surgical procedures. The mainstay of surgical treatment was untethering and spinal cord release. Section cutting of the thickened fatty filum terminale and release of the cord were performed in 58/109 neonates. The filum was found to be constricted in dorsal locations in all cases and was often accompanied by excess lipomatous material. Ten neonates underwent untethering of the spinal cord that also contained lipomyelomeningocele. Nonadhesion intradural lipomas were resected completely. Complete resection of the lipoma was not attempted if the lipoma was adherent to the spinal cord. Resection of the dermal sinus tract and release of continued adhesions to the spinal cord were carried out in 30/109 neonates. Eleven diastematomyelia neonates underwent an untethering surgical procedure. Resection of bone spurs occurred in 7 of these 11 patients (fig. 3) and resection of fibrous spurs occurred in 1 of these patients. The dural sleeve adjoining the septum was resected, and the dural sac was reconstructed. Diplomyelia without a spur was found in 3/11 patients.

Follow-Up Results and Outcome

Postoperative follow-up periods in the 109 patients, who clearly met the study criteria, ranged from 3 to 24 months (mean 12.8 months). Many patients were lost to

Table 4. Comparison of follow-up and outcomes of 171 spinal dysraphism patients with and without tethered cord syndrome

Spinal dysraphism (n = 171)	Surgery	Complication	Retethering or late tethering	Second surgery	Outcome
With tethered cord syndrome (n = 119)					
109[1]	yes	8 patients: 2 CSF leakage, 1 pseudomeningocele, 5 infection	yes (12 patients)	12 patients	1 month: 30 (28%) immediate improvement, over 3 months: 56 (51%) gradual steady improvement, after 1 year: 15 (14%) small improvement
10 (required operation, but their parents did not accept)	no	–	–	–	1 no change, 9 worse
Without tethered cord syndrome (n = 52)					
45	no	–	no	no	no change
7	no	–	yes (7 patients)	no	6 months followed: 1 worse, 6 unchanged

The patients were followed up from 3 to 48 months.
[1] Among the 109 patients who underwent an operation, 48 were followed up every 3 months from 3 to 48 months.

follow-up examinations after 24 months. Among the postoperative group, 48 patients were followed up every 3 months from 3 to 48 months. Therefore, for the reliability of the study, results included evaluations of the surgical outcomes of 109 patients before 24 months, while the results of the 48 patients examined from 24 to 48 months were studied separately.

Postoperative neurological and uroneurological evaluations of 109 patients revealed a variety of symptoms and findings. Improvements were manifested on different dates during the follow-up period. Nearly all patients (101/109, 92%) experienced improvement by 3 months. Resolution of lower-extremity weakness and sensory disturbances was seen. We found that 30/109 (28%) patients had immediate improvements as early as the first postoperative month, while in 56/109 (51%) patients, a gradual steady improvement was observed over 3 months and at 1 year. Fifteen of 109 (14%) patients had some small improvements after the 1-year follow-up. We succeeded in following 48 patients for up to 4 years. There were no significant changes between 24 and 48 months. In addition, there were no complications during surgery such as excessive bleeding or direct surgical damage to the nerve root or spine. Eight of 109 (7%) children had early postoperative complications, occurring from a few days to a month postoperatively. Two patients had cerebrospinal fluid leakage on the 2nd and 3rd postoperative days. Subcutaneous pseudomeningocele occurred in 1 patient in the first month postoperatively. Infection was observed in 5 patients. Two patients had wound infection and skin necrosis on the 4th and 15th postoperative days. Their wounds were repaired by reconstructive and cosmetic skin surgery. Meningitis was seen in 1 patient on the 2nd postoperative day though cerebrospinal fluid and blood analyses did not confirm infection. This patient recovered after 7 days and in all likelihood, it was transient viral meningitis. Paraparesis and bilateral hypernephrosis appeared in 1 patient on the 2nd postoperative day probably due to cord ischemia, which was a result of indirect surgical trauma causing neurological and uroneurological dysfunctions.

Resolution of voiding difficulty, filling phase and urinary retention was seen in 76/109 (70%) patients, and urodynamic improvements were also observed in 59/109 (54%) cases.

Nine of the 10 patients who did not undergo surgery, despite the indication for surgical treatment, showed worsening neurological and uroneurological symptoms. Only 1 child showed unchanged neurological symptoms. Urodynamic deterioration was shown in all 10 patients.

Complications, changes after surgery and outcomes of 171 patients with spinal dysraphism with or without tethered cord syndrome are summarized in table 4.

Neurological assessment was intact in 52 of 171 children with spinal dysraphism disorders without associated tethered cord syndrome; however, urinary dysfunction including frequent urination, and urinary infection developed in 7/52 (13%) patients between 3 and 9 months of age. In these 7 patients, urodynamic assessment determined the presence of a neurogenic bladder. MRI showed the occurrence of a tethered cord, although there was no initial clinical evidence of tethered cord syndrome at the first examination of these patients. All patients were followed up to 6 months and were treated conservatively. Surgical treatment was recommended to 1 patient, but her parents did not accept.

Twenty-one patients (21/109, 19%) showed a significant deterioration in neurological presentation at 6–18 months, most notably in urgency/frequency and urinary infection, whereas they had showed improvement previously. Motor weakness was present 12–18 months after surgery, probably due to retethering of the spinal cord, where an MRI examination demonstrated postoperative soft-tissue granulation causing adhesion between the cord spine and dura mater. In all patients with retethering of the spinal cord, the conus medullaris was at a low-lying level. A second untethering surgery was performed in 12 of these 21 patients. Postoperative follow-up examinations revealed improvement in 10/12 patients within 6–24 months. Two patients had a worsening of symptoms postoperatively, and both of them experienced little improvement in motor weakness by 12 months. Nine of the 21 patients showing deterioration did not undergo surgery, and they were followed up to 18 months. Moderate improvement was seen in 2 patients, while the remaining patients worsened.

Discussion

Spinal dysraphism is caused by a failure of the neural tube to close spontaneously during the 4th week of embryological development [18, 19]. This is often observed by skin markers that may not be associated with any clinical findings [19]. The cutaneous lesions indicate underlying nervous system lesions which are not apparent and are revealed only by the onset of neurological deterioration [6, 7, 11, 20–22]. Despite these findings, midline cutaneous lumbosacral lesions are not always a sign of closed spinal dysraphism [23, 24].

In the present study, we retrospectively reviewed 612 neonates with cutaneous markers among 10,000 consecutive live births. Upon review of the literature, we realized that the current study was one of the largest series among comparable studies. Kriss and Desai [12] and Kriss et al. [25] prospectively examined full-term neonates with dorsal cutaneous lesions, and each neonate underwent spinal sonography and clinical assessment of the cutaneous lesions. The incidence of cutaneous lesions in the healthy neonate study was 4.8% in the aforementioned series. Gibson et al. [7] reported on 95 neonates (1.9% of 4,989 live births) who were referred with skin markers indicating closed spinal dysraphism. In our series, we observed that the incidence of cutaneous lesions in the entire neonate study population was 6.12%. However, the incidence of closed spinal dysraphism associated with dysraphism abnormalities diagnosed with USG and MRI examinations was 1.71% in all live-born neonates and 27.94% in all neonates with skin lesions. While other studies have reported the rate to be between 20 and 30% [15], our incidence was below the maximum published incidence. We think the higher number of neonates (larger series) used in our study accounts for this difference. Other important factors that could have affected this result were the financial, social, economic, educational and intellectual level of the newborns' families. The rate of spinal-dysraphism-related nervous system abnormalities in neonates with a single and small lesions is low [26]; however, the study of 171 neonates who had spinal dysraphism abnormalities showed that multiple skin lesions, large subcutaneous masses and cutaneous sinuses over the sacrum were correlated with an increase in the rate of spinal dysraphism disorders. Henriques et al. [10] also prospectively studied minor skin lesions as signs of spinal dysraphism by spinal USG. If the ultrasound study was abnormal or inconclusive, it was complemented with MRI. Their study showed that the incidence of cutaneous lesions in all live neonates was 7.16%. This result is a little higher than our result of 6.12%.

Many authors have reported that a human tail is an infrequent cause of a tethered cord, and in the literature, there are 18 case reports [27–32]. Certainly, it is known that a dorsal cutaneous appendage (called a human tail) is often considered to be a skin marker of underlying spinal dysraphism. However, the cutaneous appendage is considered to be just a sign of spinal dysraphism and some authors have even considered these to be benign lesions without any cord abnormalities. In our study, the tail was removed, but in spite of similar causes, we did not find any relation between the tail and the spinal cord. Donovan and Pederson [27] also reported a case of human tail with noncontiguous intraspinal and spinal cord tethering. Similar to the present case, pathological ex-

amination showed some nerve fibers in the tail indicating a pseudotail though there were many nerve fibers in the pseudotail that made no contact with the spinal cord.

Due to the high incidence of severe delayed neurological problems in the newborn with closed spinal dysraphism, it is recommended that all suspected cases be evaluated. A review of the literature shows that sonography is the advised method of diagnosis. Hughes et al. [33] reported that the agreement between USG and MRI was good, particularly for the determination of a low-lying cord. They determined that 40% of USG examinations showed full agreement with MRI, 47% had partial agreement and 13% had no agreement [33]. Robinson et al. [14] investigated the rationale behind the concept that the USG was useful in screening for dysraphism in infants with cutaneous markers such as a sacral dimple or hairy patch. They concluded that as an isolated abnormality, simple dimples or pits are not useful markers of spinal dysraphism. They suggested a new imaging protocol, resulting in improved diagnostic efficiency. Allen et al. [6] performed USG and/or MRI in infants with spinal strawberry nevi. They investigated in a prospective study using a larger number of patients with isolated strawberry nevi, who underwent MRI evaluation, whether or not neuroimaging screening in these patients is indicated [6]. Babcook [34] studied the role of USG in the evaluation of infants with spinal dysraphism, but it was not the most reliable since the advent of MRI. He reported the utility of USG in evaluating neonatal spinal dysraphism as an ideal diagnostic modality for this patient population. However, comparative studies with MRI are necessary to clarify USG results. In our series, the discordance between USG and MRI was 16.58%. The mean incidence of agreement between USG and MRI was 83.42%, a little below previous studies in the literature. Consequently, the sparse clinical literature on this topic, which we reviewed, confirms that the practical attributes and financial benefits of USG recommend it as a first examination method. Although MRI is more expensive and less commonly used than USG, MRI is the most reliable examination method for the diagnosis of closed spinal dysraphism disorders, especially in cases of isolated cutaneous lesions. It is the safest technique, especially in questionable cases of spinal disorders. It is also helpful for the diagnosis of other neuropathological conditions and diseases, as well as a good guide for the management of surgical treatment. Correlation of cutaneous markers with different abnormalities by means of detailed physical assessment and radiological examinations, including USG and particularly MRI scans, makes early diagnosis and prompt treatment possible.

As was described above, a tethered cord presented with a low-lying conus medullaris on the MRI and neurological deficits upon clinical assessment. The other exceptional feature of spinal dysraphism is an anatomically normal conus medullaris with clinical findings of tethered cord syndrome, which was reported by Khoury et al. [35] in 1990. They studied a case series of 31 children who presented with uroneurological dysfunctions, where radiological examinations showed spinal dysraphism with the conus medullaris in a normal position. They stated that patients did not improve with conservative treatment and that 70% of the patients showed postoperative improvement. Steinbok et al. [36] reported that the vast majority of respondents (85%) favored surgical untethering. They also described 2 cases of disagreement among physicians, one regarding the diagnosis and treatment of a patient with symptoms of tethered cord syndrome, and the other over treatment of a patient with symptoms of tethered cord syndrome and an inconclusive MRI. They concluded that the results of the survey supported the development of a randomized clinical trial to address the benefit of surgery for tethered cord syndrome. We also presented 4 patients that had clinical evidence of a tethered cord with normal MRI findings. There was no significant difference between 1 patient who underwent surgery and 2 patients who were managed conservatively; all 3 showed minimal improvement. We found a few similar reports in the literature in which some authors clearly favored surgery [4, 37], whereas others believed that the patient did not meet the diagnostic criteria for tethered cord syndrome and that, without conclusive clinical evidence, the arguments supporting surgery for tethered cord syndrome must be viewed carefully [36, 38]. We believe this exceptional situation needs more investigations.

The management of an infant with a closed spinal dysraphism can be summarized as the early detection and treatment of a tethered cord syndrome, the prevention of further deterioration and the treatment of associated malformations [4].

Although urodynamic evaluation in neonates is difficult, it can show early physiological involvement of the spinal cord, which assists in the early diagnosis. More notably, bladder compliance is important in assessing bladder function. It helps explain the physiology that may underlie particular voiding symptoms and may reveal certain risk factors that, if unrecognized and untreated, will probably progress and cause significant urological

complications [39]. Bladder compliance may also be used to determine the postoperative physiology of the cord, particularly retethering. The long-term rate of postoperative retethering ranges from 10 to 20% [40, 41]. In our series, this value was 19% though that was over a shorter period of time, but it is still in agreement with the current literature. Retethering is not always immediately clinically significant. Postoperatively, urodynamic studies were useful in the early diagnosis of spinal cord retethering [42].

In tethered cords, the major aim of surgery is untethering and detachment of the spinal cord from the adhesive dural or paravertebral soft tissues. Detachment and removal of soft tissues should be gentle and even partial in lipomyelomeningoceles. Partial removal of subpial lipomas is performed if the lipoma in the spinal canal is bulky and does not allow for sufficient subarachnoid space around the caudal spinal cord [43]. Wehby et al. [44] reported a retrospective review of a consecutive series of 60 children with tethered cord syndrome. Postoperative outcomes included complete resolution in 52%, marked improvement in 35%, moderate improvement in 6%, minimal improvement in 6% and no improvement in 2% of children [44]. In the current study, clinical improvements in urinary symptoms occurred in 75% of patients, while 92% of the patients had experienced improvement in all neurological symptoms.

In conclusion, when spina bifida is associated with lumbar skin lesions, there may be a slightly increased incidence of coexisting tethered cord and other spinal disorders. Correlation between lumbar skin lesions and other findings can help clinicians weigh their diagnostic considerations. Careful neurological assessment and MRI examination allow for early diagnosis and prompt treatment. Urinary symptoms as a sole initial complaint and subsequent urodynamic assessments aid in early diagnosis, most notably in postoperative follow-up periods. Untethering of spinal cords to arrest deterioration is not always successful. For the best result, this population needs early and long-term follow-up. In addition, the controversy surrounding indications for surgery should be resolved, especially in cases with normal or almost normal MRI and with simple low-back dimples. The other principal point of this paper is that in neonates without skin markers, there is no disagreement for patient management, but in neonates with skin lesions with spinal disorders, sometimes the USG does not show a dysraphism and therefore may be considered as normal. Thus, MRI is usually the preferred method to evaluate neonates with dysraphism and underlying spinal disorders.

References

1 Kanev PM, Bierbauer KS: Reflections on the natural history of lipomyelomeningocele. Pediatr Neurosurg 1995;22:137–140.
2 Warder DE, Oakes WJ: Tethered cord syndrome and the conus in a normal position. Neurosurgery 1994;33:596–600.
3 Kumar R, Singh SN: Spinal dysraphism: trends in Northern India. Pediatr Neurosurg 2003;38:133–145.
4 Cornette IUC, Verpoorten C, Lagae L, Plets C, Calenbergh FV, Casaer P: Closed spinal dysraphism: a review on diagnosis and treatment in infancy. Eur J Pediatr Neurol 1998; 2:179–185.
5 Schrop P, Sorensen N, Collmann H, Krauss J: Cutaneous lesions in occult spinal dysraphism: correlation with intraspinal findings. Childs Nerv Syst 2006;22:125–131.
6 Allen RM, Sandquist AM, Piatt JH Jr, Selden NR: Ultrasonographic screening in infants with isolated spinal strawberry nevi. J Neurosurg 2003;98(Spine suppl 3):247–250.
7 Gibson PJ, Britton J, Hall DM, Hill CR: Lumbosacral skin markers and identification of occult spinal dysraphism in neonates. Acta Pediatr 1995;84:208–209.
8 Soonawala N, Overweg-Plandsoen WC, Brouwer OF: Early clinical signs and symptoms in occult spinal dysraphism: a retrospective case study of 47 patients. Clin Neurol Neurosurg 1999;101:1–4.
9 Shian WJ, Chi CS, Wong TT: Lipomyelomeningiocele: a nine year review. Acta Pediatr Sin 1994;35:57–62.
10 Henriques JG, Pianetti G, Henriques KS, Costa P, Gusmao S: Minor skin lesions as markers of occult spinal dysraphisms – prospective study. Surg Neurol 2005;63(suppl 1):8–12.
11 Sattar MT, Bannister CM, Turnbull IW: Occult spinal dysraphism: the common combination of lesions and the clinical manifestations in 50 patients. Eur J Pediatr Surg 1996; 6:10–14.
12 Kriss VM, Desai NS: Occult spinal dysraphism in neonates: assessment of high-risk cutaneous stigma on sonography. AJR Am J Roentgenol 1998;171:1987–1992.
13 Pierre-Kahn A, Zerah M, Renier D, Cinalli G, Sainte-Rose C, Lellouch-Tubiana A, Brunelle F, Le Merrer M, Giudicelli Y, Pichon J, Kleinknecht B, Nataf F: Congenital lumbosacral lipomas. Child Nerv Syst 1997;13:298–335.
14 Robinson AJ, Russel S, Rimmer S: The value of ultrasonic examination of the lumbar spine infants with specific reference to cutaneous markers of occult spinal dysraphism. Clin Radiol 2005;60:72–77.
15 Ward PJ, Clarke NM, Fairhurst JJ: The role of magnetic resonance imaging in the investigation of spinal dysraphism in the child with lower limb abnormality. J Pediatr Orthop B 1998;7:141–143.
16 Perra GK, Atherton D: The value of MRI in a patient with occult spinal dysraphism. Pediatr Dermatol 2006;23:24–26.
17 Santiago Medina L, Al-Orfali M, Zurakowski D, Poussaint Ty, DiCanzio J, Barnes PD: Occult lumbosacral dysraphism in children and young adults: diagnostic performance of fast screening and conventional MR imaging. Radiology 1999;211:767–771.
18 Drolet BA: Cutaneous signs of neural tube dysraphism. Pediatr Clin North Am 2000; 47:813–823.
19 Van Allen MI, Kalousek DK, Chernoff GF, Juriloff D, Harris M, McGillivray BC, Yong SL, Langlois S, MacLeod PM, Chitayat D, et al: Evidence for multisite closure of the neural tube in humans. Am J Med Genet 1993; 42:723–743.

20 Guggisberg D, Hadj-Rabia S, Viney C, Bodemer C, Brunell F, Zerah M, Kahn AP, De Prost Y, Hamel-Teillac D: Skin markers of occult spinal dysraphism in children: a review of 54 cases. Arch Dermatol 2004;140: 1109–1115.
21 Tubbs RS, Wellons JC 3rd, Iskandar BJ, Oakes WJ: Isolated flat capillary midline lumbosacral hemangiomas as indicators of occult spinal dysraphism. J Neurosurg 2004; 100(Pediatrics suppl 2):86–89.
22 Pacheco-Jacome E, Ballesteros MC, Jayakar P, Morrison G, Ragheb J, Medina LS: Occult spinal dysraphism: evidence based diagnosis and treatment. Neuroimag Clin North Am 2003;13:327–334.
23 Humphreys RP: Clinical evaluation of cutaneous lesions of the back: spinal signatures that do not go away. Clin Neurosurg 1995;43: 176–187.
24 Martinez-Lage JF, Niguez BF, Perez-Espejo MA, Almagro MJ, Maeztu C: Midline cutaneous lumbosacral lesions: not always a sign of occult spinal dysraphism. Childs Nerv Syst 2006;1:1–5.
25 Kriss VM, Kriss JC, Desai NS, Warf BC: Occult spinal dysraphism in the infant. Clin Pediatr 1995;34:650–654.
26 Sarica K, Erbagci A, Yagci F, Yurtseven C, Buyukcebeci O, Karakurum G: Multidisciplinary evaluation of occult spinal dysraphism in 47 children. Scand J Urol Nephrol 2003;37:329–334.
27 Donovan DJ, Pederson RC: Human tail with noncontiguous intraspinal lipoma and spinal cord tethering: case report and embryologic discussion. Pediatr Neurosurg 2005;41: 35–40.
28 Gonul E, Izci Y, Onguru O, Timurkaynak E, Seber N: The human tail associated with intraspinal lipoma: case report. Minim Invas Neurosurg 2000;43:215–218.
29 Muthukumar N: The 'human tail': a rare cause of tethered cord – a case report. Spine 2004;59:476–478.
30 Birol A, Bademc G: Faun tail diagnosis of occult spinal dysraphism with a rare cutaneous marker. J Dermatol 2004;31:251–252.
31 Park SH, Huh JS, Cho KH, Shin YS, Kim SH, Ahn YH, Cho KG, Yoon SH: Teratoma in human tail lipoma. Pediatr Neurosurg 2005;41: 158–161.
32 Lu FL, Wang PJ, Teng RJ, Yau KI: The human tail. Pediatr Neurol 1998;19:230–233.
33 Hughes JA, De Bruyn R, Patel K, Thompson D: Evaluation of spinal ultrasound in spinal dysraphism. Clin Radiol 2003;58:227–233.
34 Babcook CJ: Ultrasound evaluation of prenatal and neonatal spina bifida. Neurosurg Clin North Am 1995;6:203–218.
35 Khoury AE, Hendrick EB, McLorie GA, Kulkarni A, Churchill BM: Occult spinal dysraphism: clinical and urodynamic outcome after division of the filum terminale. J Urol 1990;144:426–429.
36 Steinbok P, Garton HJL, Gupta N: Occult tethered cord syndrome: a study survey of practice patterns. J Neurosurg 2006;104: 309–313.
37 Seden NR: Occult tethered cord syndrome: the case for surgery. J Neurosurg 2006; 104(pediatrics suppl 5):302–304.
38 Drake JM: Occult tethered cord syndrome: not an indication for surgery. J Neurosurg 2006;104(Pediatrics suppl 5):305–308.
39 Kaplan SA, Blaivas JG: Evaluation and management of the neurogenic bladder; in Pang D (ed): Disorders of the Pediatric Spine. New York, Raven Press, 1995, pp 87–95.
40 Colak A, Pollack IF, Albright AL: Recurrent tethering: a common long-term problem after lipomyelomeningocele repair. Pediatr Neurosurg 1998;29:184–190.
41 Houser EE, Bartholomew TH, Cookson MS, Marlin AE, Little NA: A prospective evaluation of leak point pressure, bladder compliance and clinical status in myelodysplasia patients with tethered spinal cords. J Urol 1994;151:177–180, discussion 180–181.
42 Meyrat BJ, Tercier S, Lutz N, Rilliet B, Forcada-Guex M, Vernet O: Introduction of a urodynamic score to detect pre- and postoperative neurological deficits in children with a primary tethered cord. Childs Nerv Syst 2003;19:716–721.
43 Koyanagi L, Iwasaki Y, Hida K, Abe H, Isu T, Akino M, Aida T: Factors in neurological deterioration and role of surgical treatment in lumbosacral lipoma. Childs Nerv Syst 2000; 16:143–149.
44 Wehby MC, O'Hollaren PS, Abtin K, Hume JL, Richards BJ: Occult tight filum terminale syndrome: results of surgical untethering. Pediatr Neurosurg 2005;41:58, author reply 59–60.