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Tethered cord syndrome

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Burth S, Berger TM, Winiker H, Caduff JH, Schmitt-Mechelke T, Kothbauer KF, Neonatal and Pediatric Intensive Care Unit (BS, BTM), Department of Pediatic Surgery (WH), Department of Pediatric Radiology (CJH), Department of Neuropediatrics (SMT), Children's Hospital of Lucerne, Department of Neurosurgery (KFK), Cantonal Hospital of Lucerne, Switzerland

This female patient was born to a 40-year-old G4/P2 at term by elective caesarean section because of breech presentation. Pregnancy had been uneventful. Maternal serologies (HBV, HCV, HIV, lues, rubella and toxoplasmosis) as well as prenatal routine ultrasound scans had been unremarkable and there was no history of illicit drug use, nicotine or alcohol consumption. The father of the child has type 1 diabetes mellitus. The two-year-old brother is healthy. There is no family history of congenital malformations.

The baby girl adapted well with normal Apgar scores. Her birth weight, length and head circumference were appropriate for gestational age (all between P50 and P75). On examination shortly after birth, she was noticed to have a mass over the lumbosacral region and a sacral dimple approximately 1 cm below this mass (Fig. 1). The remainder of her physical examination was normal.

A spinal ultrasound revealed an occult sacral spina bifida, a low-lying medullary conus at the level of L5/S1, a tethered cord as well as a lipoma within the sacral canal (Fig. 2). An intraspinal extension of the dermal sinus could not be ruled out. Even on detailed neurological examination, muscular tone, deep tendon reflexes (DTRs), anal reflex and sensibility of the lower extremities appeared to be normal. Voiding was unimpaired without residuals after micturition.

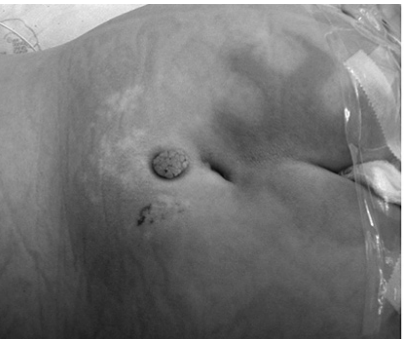
At 3 months of age, a spinal MRI confirmed the low-lying conus with the apex at the level of S1/S2, an intraspinal lipoma extending from S1 caudally to the right and dorsally of the conus and the filum terminale, and a tethered cord (Fig. 3). All sacral vertebrae were bifid, and there was a sacro-cutaneous duct or tract and a blindly ending duct beginning over the tip of the coccyx. Cranial MR images were normal.

Clinically, the girl was thriving and her neurological examination remained without abnormality. The parents reported normal voiding and bowel movements. They had the impression that their daughter might be less sensitive to painful stimuli than one might expect. A voiding cystourethrogram (VCUG) revealed no reflux but signs consistent with a neurogenic bladder. The family was informed about the concept of prophylactic untethering in the first year of life. Some evidence of incipient neurogenic bladder and the uncertainty about the patency of the sacrocutaneous duct representing an infection risk added to the argument for surgery.

At nine months of age, she was admitted for the operation. At that time, the deep tendon reflexes appeared slightly reduced but otherwise the neurological examination continued to be unremarkable. At surgery, the tethered conus and the lipoma were exposed. Untethering was accomplished through partial lipoma resection and removal of all fibrous attachment of

the conus to the overlying tissue. The resulting dorsal dural defect was closed with a synthetic graft, and the wound was tightly closed using a standard technique (1) (Fig. 4, 5). The post-operative course was complicated by a urinary tract infection caused by an extended-spectrum ß-lactamase (ESBL) producing E. coli and a local wound infection associated with the same agent resulting in a CSF leak that required neurosurgical revision and reclosure as well as transient treatment with acetazolamide. Her further hospital course was unremarkable and she was discharged home two weeks later. At the age of 12 months, there are no apparent neurological deficits except for some degree of urinary and fecal incontinence.

Lumbosacral mass and sacral dimple (aspect at the time of surgery).



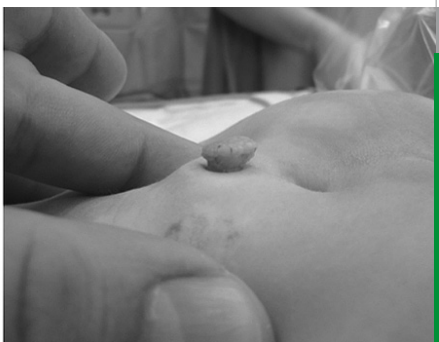


Fig. 1

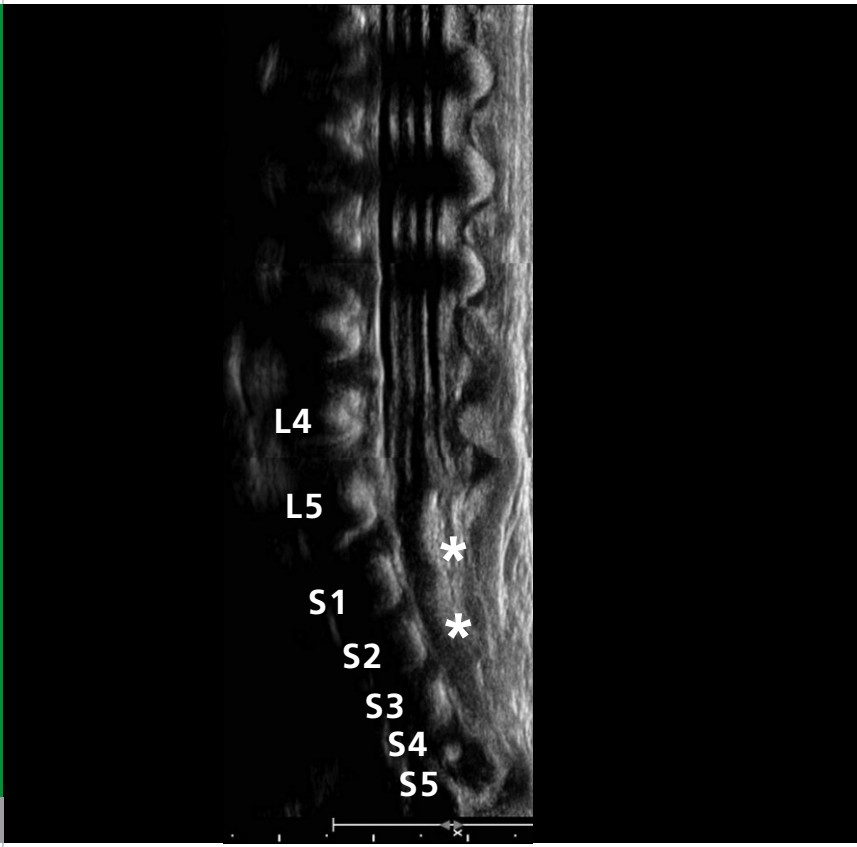


Fig. 2

Ultrasound examination (day 3 of life): occult spina bifida, low-lying medullary conus and intraspinal lipoma (asterisks).

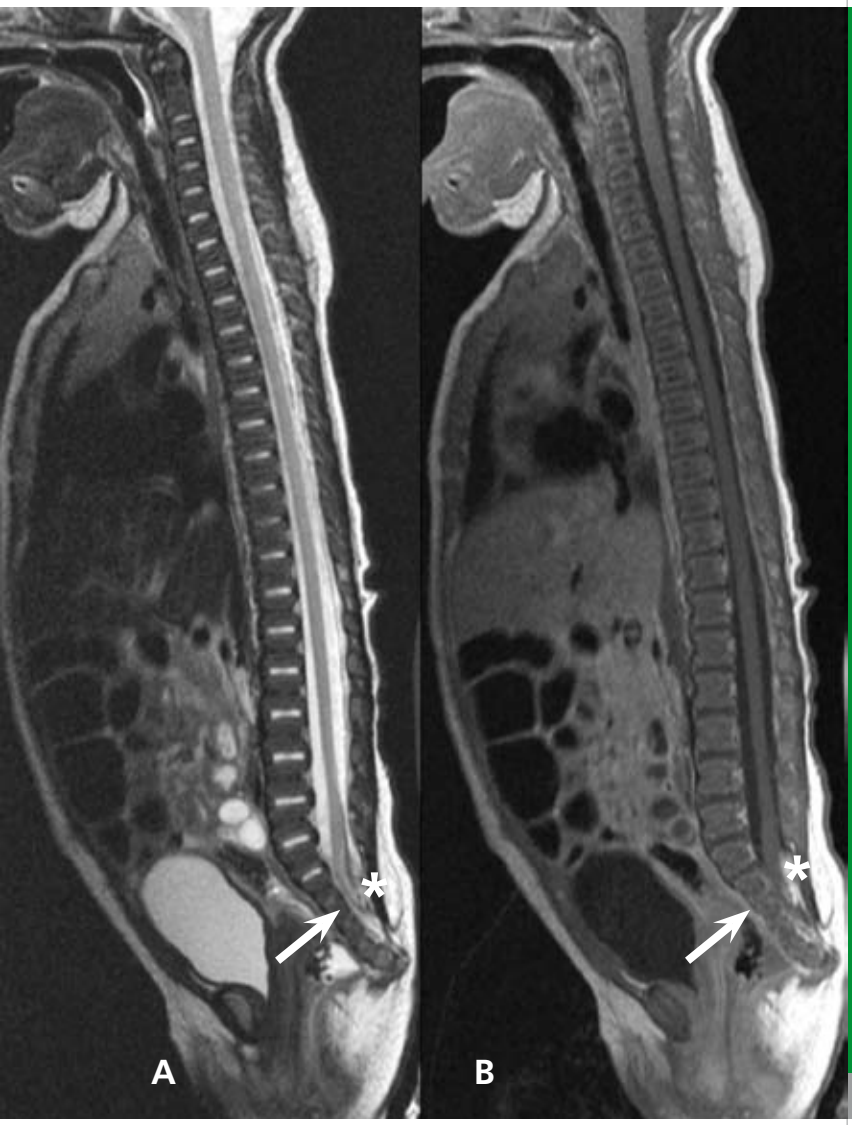


Fig. 3

MRI of the spinal canal (age 3 months): low-lying medullary conus at the level of S1/S2 (arrow) and lipoma associated with the filum terminale (asterisk) (A: T2-weighted image, B: T1-weighted image).

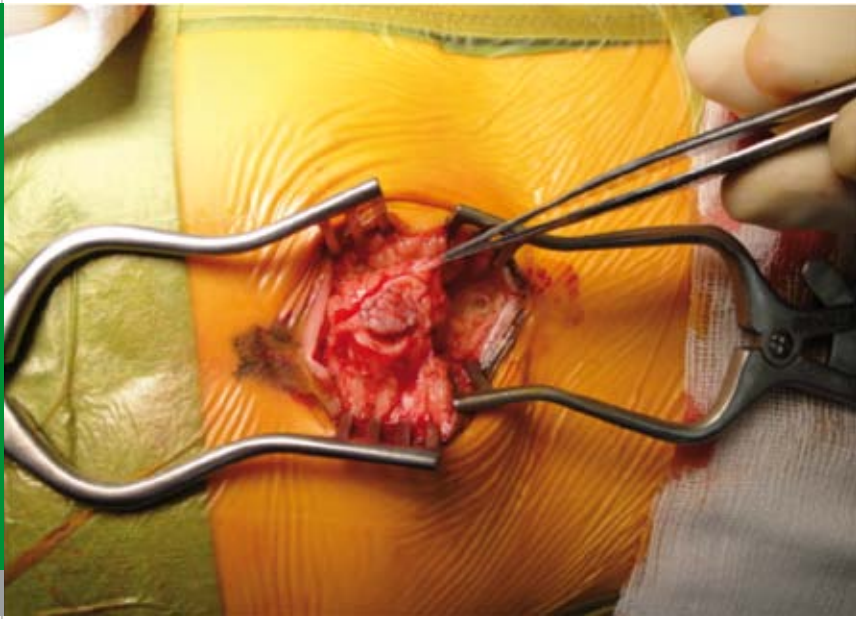


Fig. 4

Excision of the cutaneous mass.

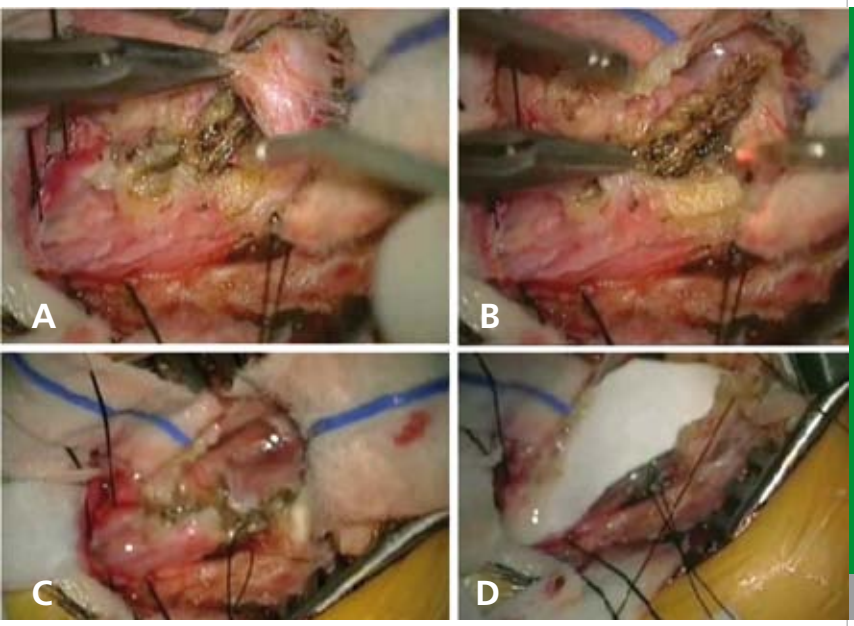


Fig. 5

Intraoperative appearance of the tethered cord: A) conus lifted with nerve roots, B) conus open and lipoma removed, C) conus sutured, D) dural graft.

DISCUSSION

Open or closed spinal dysraphism occurs with a frequency of 0.5-8:1000 live births (2). There is no unified risk factor for all occult spinal dysraphic lesions and depending on the stage of development of the nervous system at which an insult occurs one or more dysraphic lesions can develop. Folic acid deficiency has been clearly associated with open spinal dysraphism (3) but its relationship with closed spinal dysraphism has not been evaluated (4). Another well-known risk factor for spina bifida is maternal exposure to valproate or carbamazepine (5-7).

Closed spinal dysraphic anomalies can be classified into three groups based on the developmental stage at which they are thought to arise (Table). One or more dysraphic lesions commonly occur in the same patient (8).

Table

Classification of closed spinal dysraphic lesions.

Abnormalities of notochord development	▪ neurenteric cysts ▪ split notochord syndrome ▪ split spinal cord malformations ▪ sacral meningeal cysts or sacral meningocele ▪ dorsal dermal sinus tracts and cysts
Abnormalities of primary neurulation	▪ syringohydromyelia ▪ spina bifida occulta ▪ spinal lipomas and teratomas
Anomalies of the caudal cell mass and secondary neurulation	▪ tethered cord syndrome (TCS) ▪ terminal diplomyelia ▪ sacrococcygeal teratomas ▪ caudal regression or sacral agenesis

Failure of fusion of the posterior vertebral arches during primary neurulation results in spina bifida occulta. Minor abnormalities of the overlying skin (naevi, dermal sinus or dimple, underlying lipoma or a hirsute area) are common (9).

Spinal lipomas are less common than spinal dysraphic lesions (1:4000 live births) (10). They are thought to arise from abnormal mesodermal cells that failed to migrate normally and hence got trapped between the roof plate and the ectoderm. They often contain various types of tissue and thus can be considered complex teratomas (11). They may compress the cord and thereby cause progressive neurological dysfunction. Most patients with spinal lipomas have cutaneous stigmata (subcutaneous lump or dermal sinus). Spinal lipomas frequently coexist with a tethered cord in the same patient.

Tethered cord syndrome (TCS) is a stretch-induced dysfunction of the caudal spinal cord and conus. Normally, the filum terminale is elastic and does not cause traction on the spinal cord during inclination. In TCS, the filum terminale is caudally attached to inelastic structures causing a low-impact high frequency stretch injury upon the cord (12). This may cause repeated microtrauma, which in the long run leads to first neuronal and later axonal degeneration subsequently resulting in irreversible neurological deficits (13, 14).

The diagnosis of closed spinal dysraphism in a neonate requires careful clinical examination. Symptoms leading to the diagnosis can be cutaneous abnormalities, upper or lower motor neuron dysfunction and associated malformations. Since detection of subtle neurological disturbances and clinical evaluation of bladder function are obviously difficult in a neonate, typically located cutaneous stigmata (present in about 80% of cases) are of great diagnostic value (14). Meticulous clinical examination of the skin all along the spine is mandatory. Ultrasonography of the spinal canal is the initial investigation of choice in an asymptomatic infant with cutaneous stigma in the lumbosacral region. At about three months of age, an MRI of the spine and the brain should be obtained for a definitive anatomical diagnosis. Urological assessment during the first 3 months of life is mandatory and should include microscopic urinalysis, periodic urine cultures, renal function tests, a VCUG and ultrasonography of the bladder and the kidneys.

Key elements of successful management of an infant with closed spinal dysraphism are early detection and treatment of a TCS, prevention of further deterioration and treatment of associated malformations. A multidisciplinary team approach involving pediatric neurologists, pediatric neurosurgeons, urologists, orthopedic surgeons, neonatologists, physiotherapists and social workers is required. Indication for and timing of surgical untethering in patients with TCS continues to

be a controversial issue between the here-proposed prophylactic concept and a conservative wait-and-see approach (15, 16). The main principles of surgery are complete untethering and dural reconstruction with adequate CSF space around the spinal cord to prevent retethering (17).

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Swiss Society of Neonatology
www.neonet.ch
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