Rehabilitation of a secondary paraplegia due to diastematomyelia: Case report

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Introduction.— Diastematomyelia is a rare spinal cord malformation involving a division of the cord in the sagittal plan giving rise to two hemi-cords leading to central nervous system disorders requiring surgical treatment and adapted rehabilitation.

Observation.— We reported the case of a 23-year-old girl with an uneventful history who complained of back pain for one month associated with urine loss, paraesthesias and gradually a total functional impotency of both lower limbs. The physical examination revealed the presence of a dorsal hyperpilosity and right dorso-lumbar scoliosis with a flank neurological examination. The cord MRI showed a diastematomyelia standard II with a centromedullaire ossified spur located at T9–T10. T9–T10 laminectomy was performed with excision of an osseous spur. The postoperative examination noted: absence of improvement of the paraesthesia with a pyramidal syndrome, a posterior cord syndrome and a neuro-bladder. During hospitalization in our unit, the patient participated in twice-daily sessions of functional rehabilitation. The outcome was marked by a recovery of walking ability with spontaneous micturition and complementary catheter insertion.

Discussion.— Diastematomyelia is a rare neurological malformation which accounts for 4% of all forms of dysraphisms, with clear female prevalence. As in our case, the diastematomyelia is usually thoracolombaire whereas the cervical localization is exceptional. Diastematomyelia can cause a major handicap. The diagnosis is confirmed by the MRI, the treatment of choice is surgery. Rehabilitation is crucial for diastematomyelia patients, but seldom described in the literature. Appropriate rehabilitation makes it possible to improve the functional status and quality of life of these generally young patients.


Mayer-Rokitansky-Küster-Hauser syndrome associated with spinal cord AV malformation

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Case review.— Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, actually uroterovaginal agenesis, is a congenital non-formation of the vagina and the uterus, but with normal ovaries. Its clinical features include partial or complete absence (agenesis) of the uterus with an absent or hypoplastic vagina, normal fallopian tubes, ovaries, normal external genitalia and the typical 46, XX, female chromosom pattern. Secondary sexual characteristics are present. Associated renal, heart, hearing and/or skeletal abnormalities are common. Mayer-Rokitansky-Kuster-Hauser syndrome usually remains undetected until the patient presents with primary amenorrhoea despite normal female sexual development. Female, 41 years old, with MRKH syndrome presenting non-specific, suddenly caused pain in lower limbs (more in right leg) and sudden paralysis of lower limbs with walking disability. This patient presented primary amenorrhoea, leading to a diagnosis of congenital absence of the upper vagina and uterus, with normal bilateral adnexa, and normal secondary sexual characteristics, normal karyotype (46, XX). The patient had no visceral malformations (the heart and kidneys, in particular, were normal, as assessed by ultrasound examinations) or hearing impairment. Skeletal abnormalities were observed with radiography and CT scan. Congenital malformation of vertebral canal and hemic vertebra Th 10, cleavage of the body of L4 vertebra, and scoliosis of lumbar part were confirmed.


References
