

A case of paraplegia in a child with osteogenesis imperfecta

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Introduction

Osteogenesis imperfecta (OI) is a hereditary disorder that courses with skeletal abnormalities. Although progressive spinal deformities are frequent, there are few cases of related neurological complications reported. Rehabilitation of both spinal cord injury and OI is a demanding achievement, which requires multidisciplinary efforts. The authors' aim is to report the case of a child with OI and non-traumatic spinal cord injury throughout her inpatient rehabilitation program in our department.

Case description

A 12 year old child with OI presented with progressive deterioration and loss of walking ability, urge urinary incontinence and constipation, four months prior to the admission to our hospital for further investigation. She had a past medical history of multiple fractures and learned to walk with crutches at the age of five years. Clinical examination revealed multiple skeletal deformities and a severe kyphoscoliosis affecting C6 to T9 segments. The ASIA impairment scale grade was a sensory incomplete paraplegia AIS B with T6 sensory level. Neurological examination showed hypotonia and hyperreflexia of lower limbs. She had a score of 60/126 in Functional Independence Measure (FIM) scale. Magnetic resonance imaging (MRI) of the spinal cord revealed myelomalacia and atrophy at T3 level. The urodynamics study showed a mild hyperactive bladder.

Therapy

The rehabilitation aim was to improve the child's motor function and autonomy in daily life competencies, and to minimize the disease burden. During the stay in our hospital the child had daily sessions, five days a week, of occupational and physical therapies. The child and caregivers were taught intermittent vesical catheterization technique and began to do bladder drainages every three hours during daytime. Oxybutynin was prescribed for better control of vesical symptoms. Intestinal training with precise schedule and laxatives was established. She was medicated with Baclofen for spasticity treatment. Technical aids of daily life use were studied and prescribed. The child's home had been previously adapted to provide maximum autonomy.

Outcome

By the end of the three months rehabilitation program in our department the child returned home referred to her local area hospital. At this time she presented a paraplegia ASIA C T6 level. She had better postural control and trunk balance, and an improvement in FIM scale with a 74/126 score.

Conclusions

Both conditions require a wide approach in order to reduce functional impairment. A regular follow-up by a paediatric rehabilitation specialist and long-term treatment with physical and occupational therapies should be maintained in order to decrease the occurrence of complications and to provide a healthy growing.



Figure 1 – Coronal 3D Computed tomography of vertebral column



Figure 2 – Figure 1 – Sagittal 3D Computed tomography of vertebral column

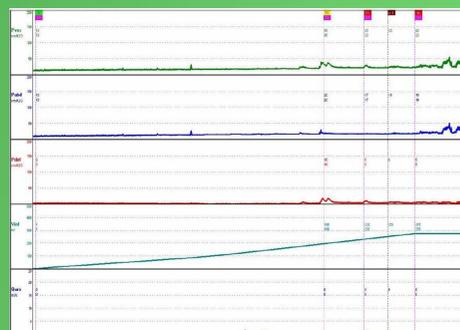


Figure 5 – Urodynamics study



Figure 6 – T2-weighted oblique sagittal MRI of thoracic spinal cord

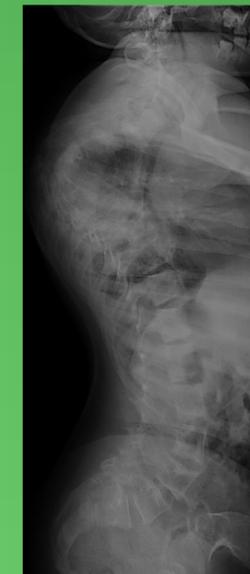


Figure 3 – Sagittal x-ray of vertebral column x



Figure 4 – Coronal x-ray of vertebral column

