Hellenic Spine Society

neuromuscular scoliosis

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neuromuscular scoliosis

- Multitude of diseases
- ALL children with type of neuromuscular disease have a tendency for scoliosis
- Severity of disease and severity of deformity

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neuromuscular scoliosis

- Long and sweeping curves, type C
- Rare compensatory curves
- Pelvic obliquity
- Involvement of cervical spine

- PULMONARY FUNCTION

neuromuscular scoliosis surgical treatment

- Long fusion area
- Osteoporotic bone
- Blood loss
- Surgery is MAINLY performed to achieve stability and balance, NOT to correct deformity

Cerebral palsy

Non progressive lesion affecting posture and gait, after a CNS lesion during pre, peri and post natal period

Cerebral palsy classification

- Tetraplegia
  - Diplegia
- Hemiplegia
- double hemiplegia

- Spasticity
- Diskinesia
- Ataxia
- mixed

General Motor Function Classification System

ON THE OTHER HAND

Carl L. Stanitski, MD, Editor

Classifying Cerebral Palsy

H. Kerr Graham, MD, FRCS(Ed), FRACS

Each year, the *Journal of Pediatric Orthopaedics* publishes more papers dealing with orthopaedic issues of children with cerebral palsy (CP) than any other medical journal. However, it can sometimes be difficult for the reader to understand the defining features of the study population. Different authors use different terms, and the definitions of the terms are imprecise. CP is traditionally classified by motor type and topographical distribution. A classification based on motor type might include the terms *spastic, dyskinetic, ataxic, hypotonic,* and *mixed.* The most commonly used terms in classifications of topographical distribution are *hemiplegia, diplegia,* and *quadriplegia,* but the terms *monoplegia, paraplegia, triplegia, double hemiplegia* and *tetraplegia* are also used. The terms vary considerably, but more importantly classifications by motor type and topography are known to be unreliable.[1]

What can contributors to JPO do to improve communication? The answer is to add a simple, valid, and reliable classification of gross motor function to their clinical

FIGURE 1. The Gross Motor Function Classification System (GMFCS) for children aged 6 to 12 years. GMFCS level I: Children walk indoors and outdoors and climb stairs without limitation. Children perform gross motor skills including running and jumping, but speed, balance, and co-ordination are impaired. GMFCS level II: Children walk indoors and outdoors and climb stairs holding onto a railing but experience

Cerebral palsy

- **Table 1**
- **Summary of the GMFCS classification**
- GMFCS LevelDescription
- I Walks without limitations. Limitations in more advanced motor skills
- II Walks without assistive devices. Limitations walking outdoors and in the community
- III Walks with assistive mobility devices. Limitations walking outdoors and in the community
- IV Self-mobility with limitations. Children are transported or use power mobility outdoors or in the community
- V Self-mobility severely limited even with the use of assistive technology

Cerebral palsy

- Incidence and severity of scoliosis depends on the level of NM involvement
- Hemiplegia Rare to have a curve >10d

- Tetraplegia non ambulants wheel chair bound, almost all of them

Cerebral palsy

- Continuous physiotherapy and standing frames are essential to delay scoliosis
- Progression of curves as the patient is growing in height and weight

- hydrotherapy

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Progression of scoliosis in Cerebral palsy

- Loss of sitting balance
- Unable to use upper limbs
- Sleeping in a fixed position

Cerebral palsy conservative management

- Anatomic seat
- Molded chair to achieve a better position

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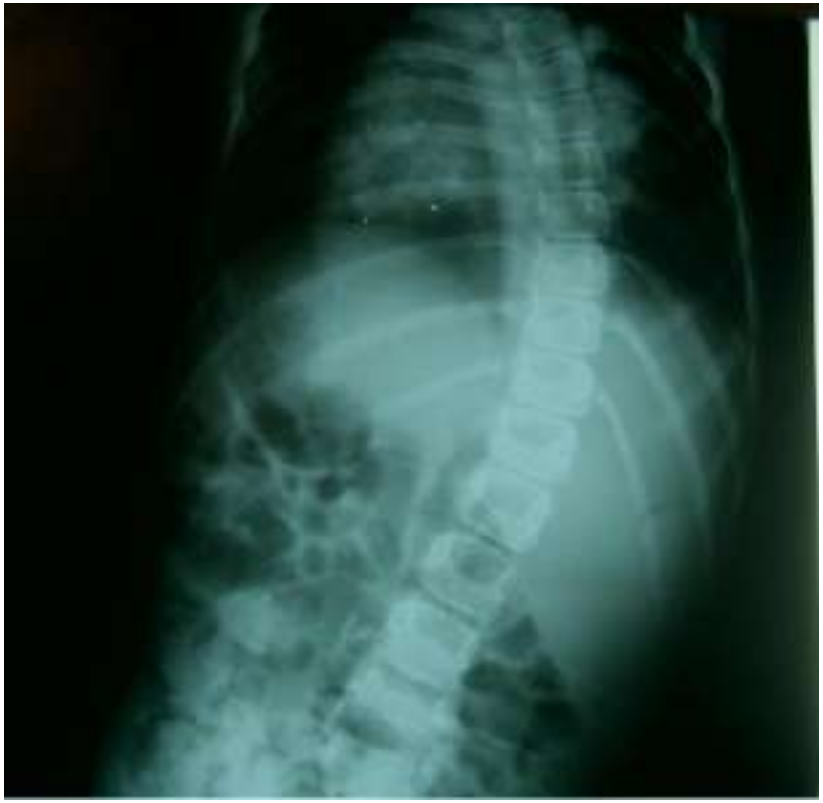
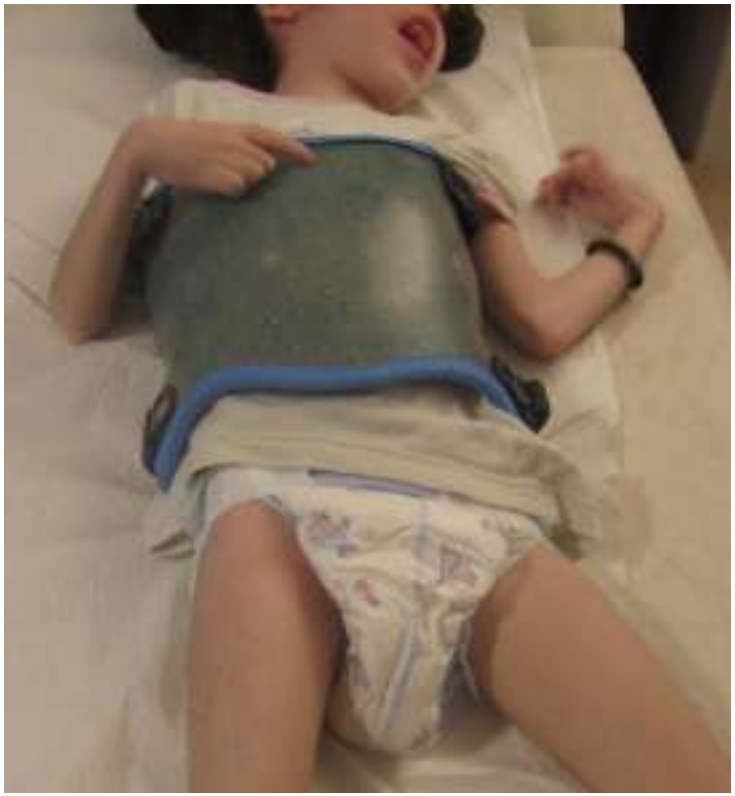
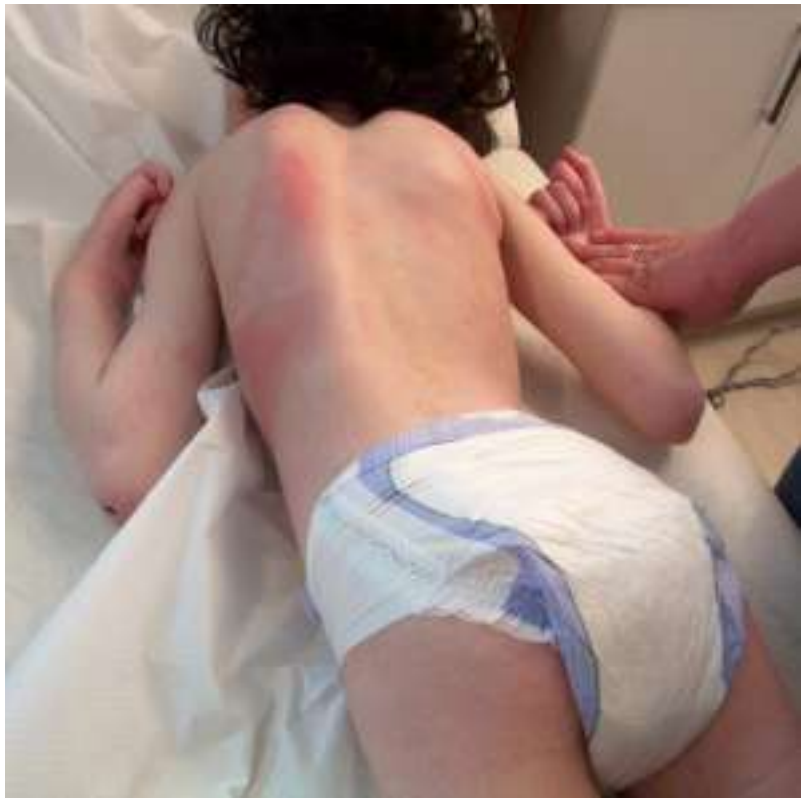
Cerebral palsy conservative management

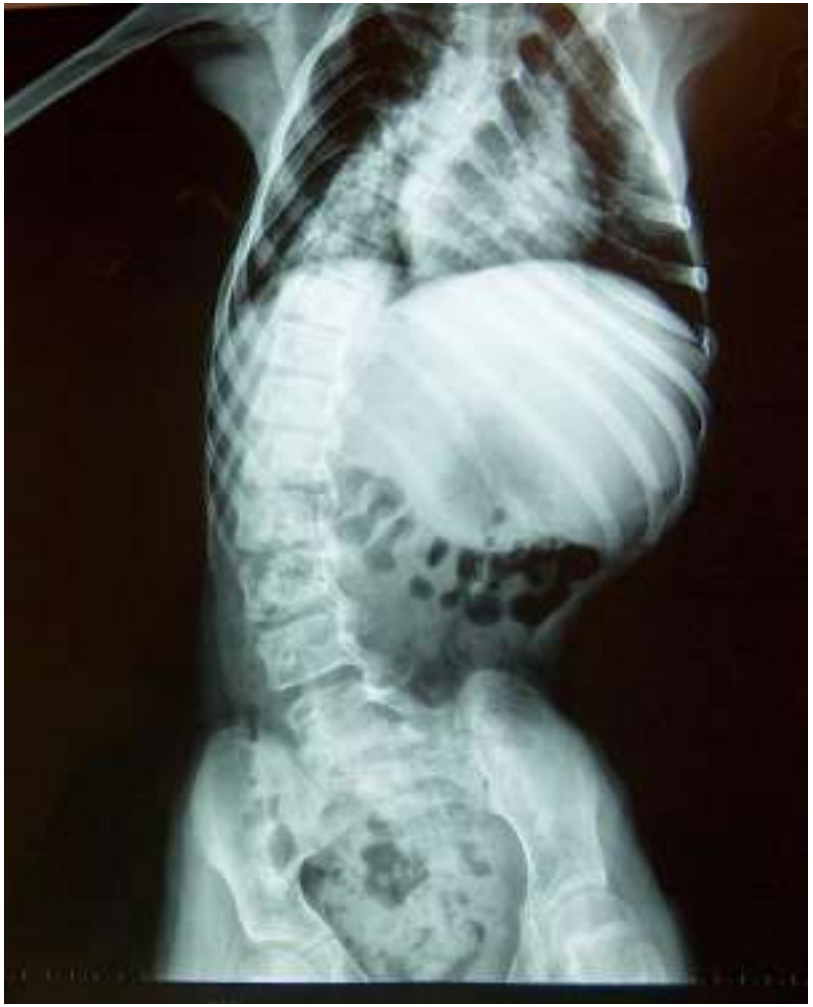




Tetraplegic CP Total involvment

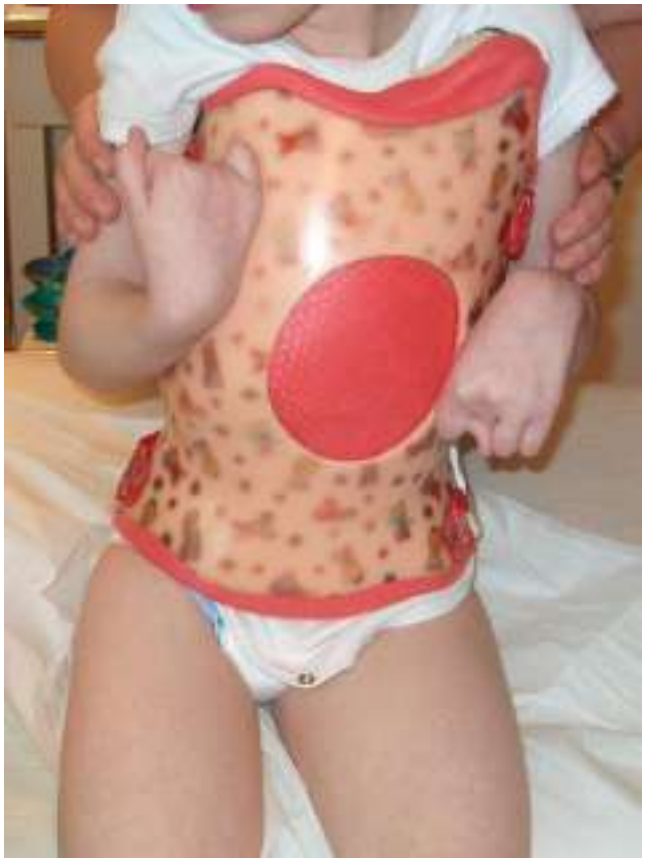
- Appropriate orthotics





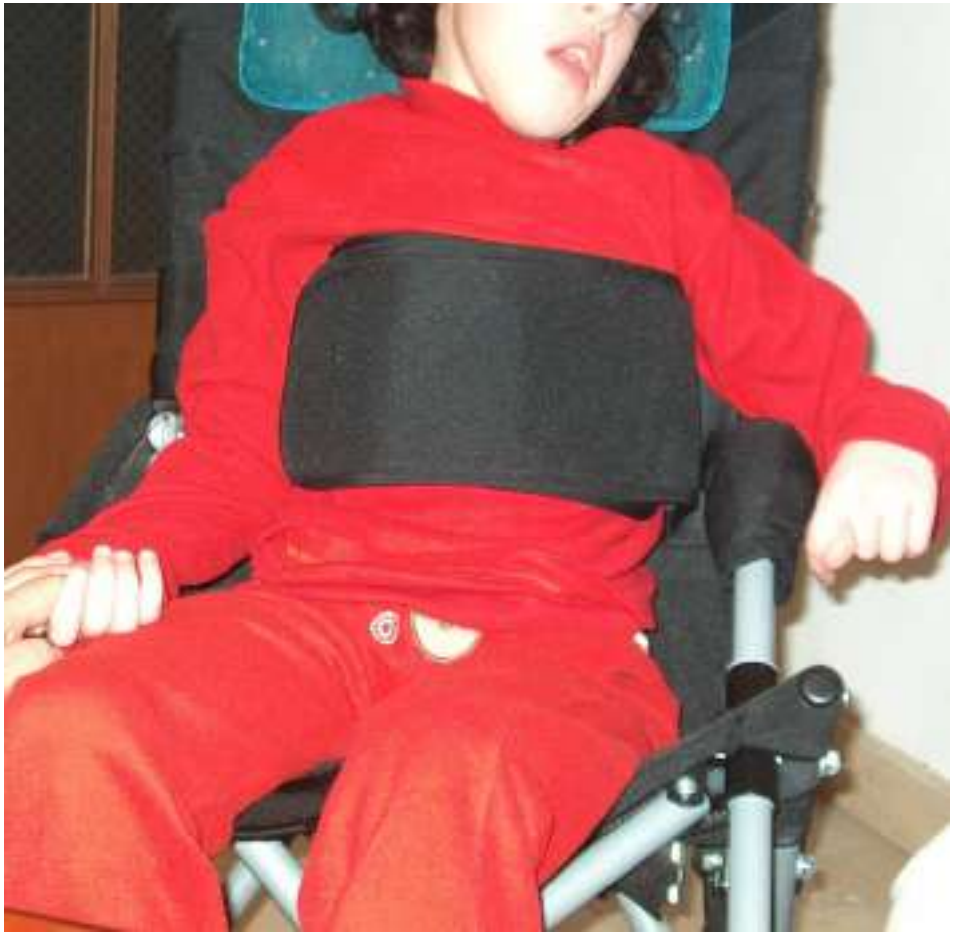
CP Total involvment

- Appropriate orthotics

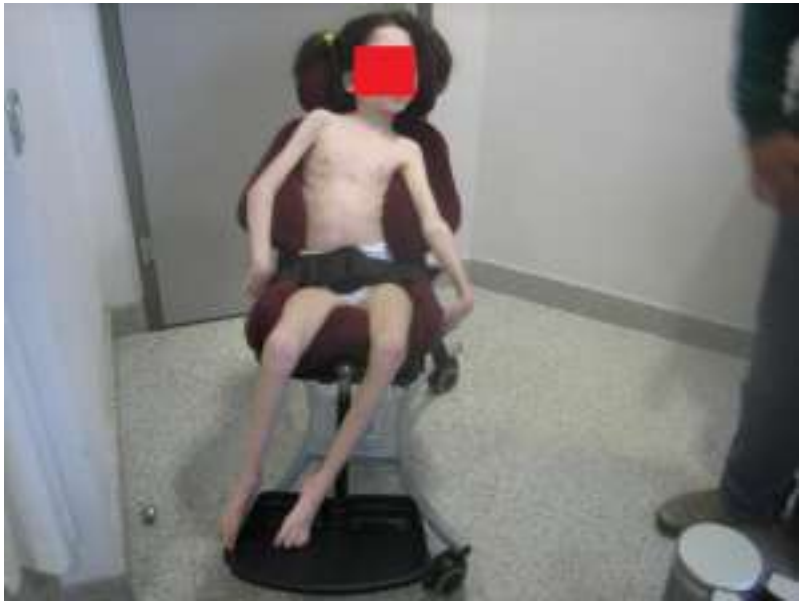


Tetraplegic CP Total involvment

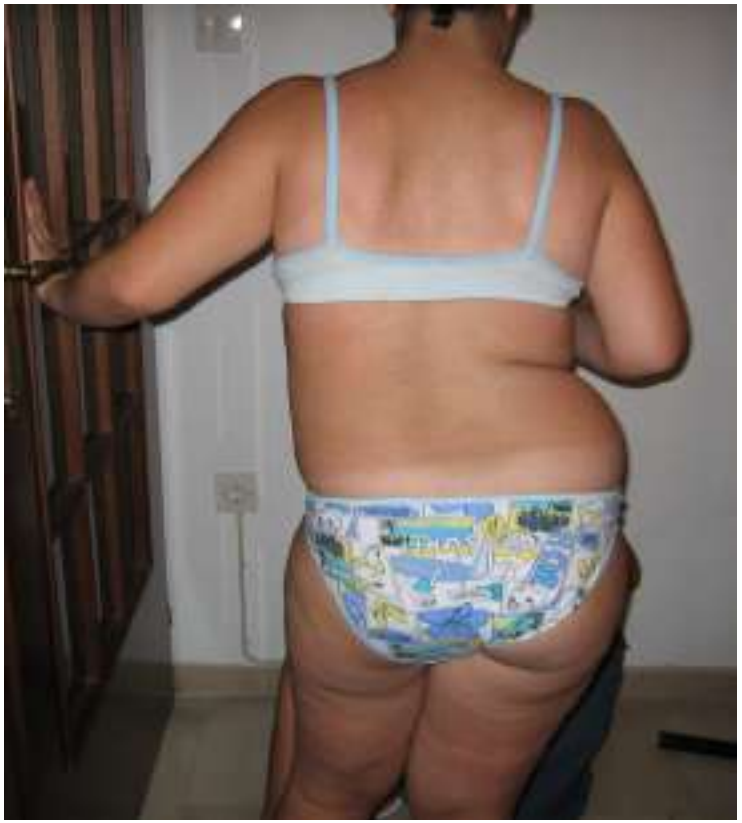
- Wheel chair seat

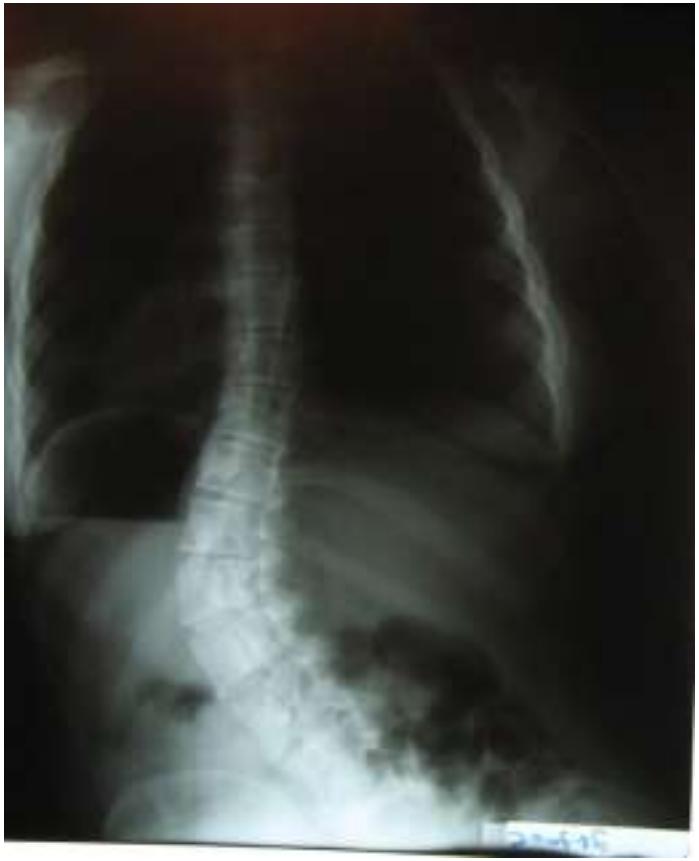


Anatomical seat body sealed



Psychomotor delay (mixed type)





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Cerebral palsy surgical treatment

- Curves of the thoracic or thoracolumbar region with parallel pelvis
- More severely involved children with pelvic obliquity

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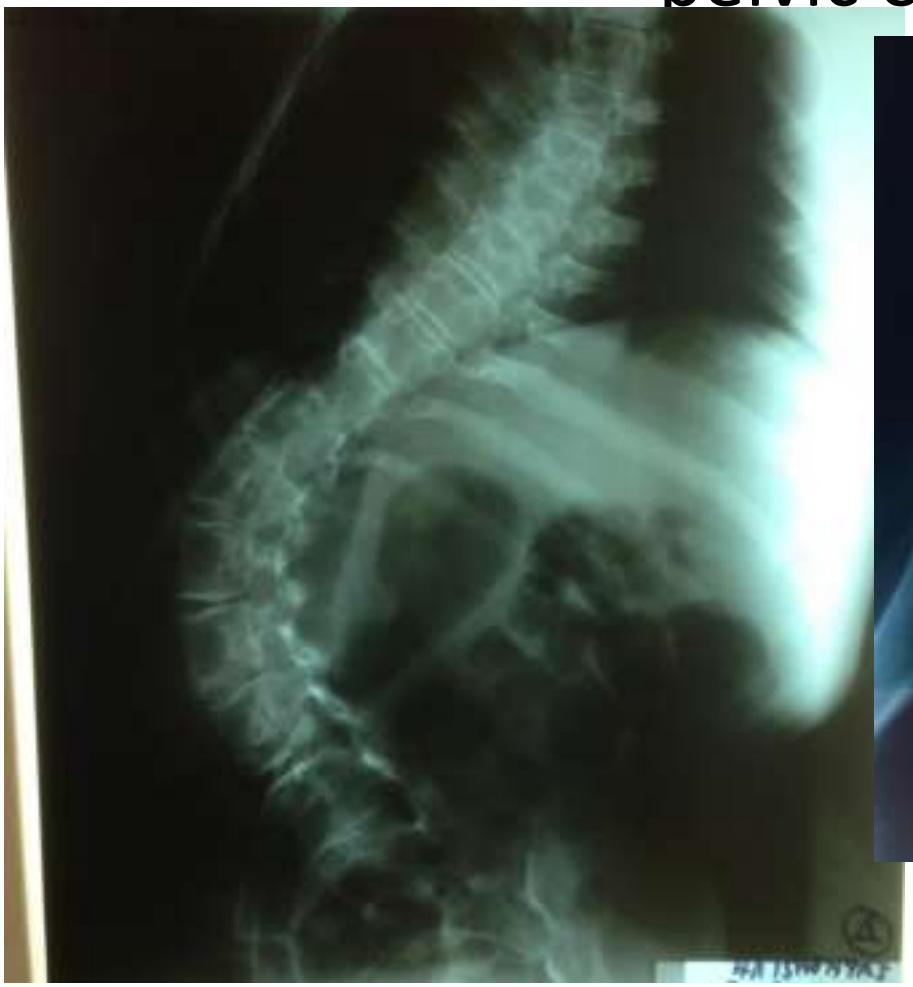
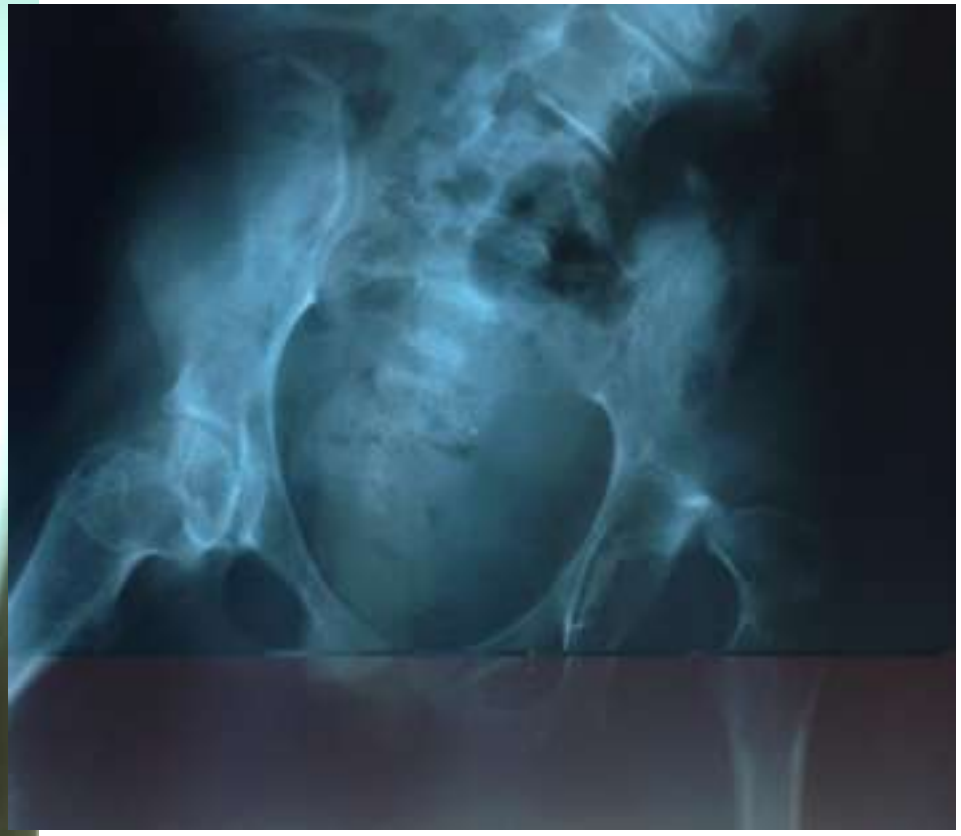
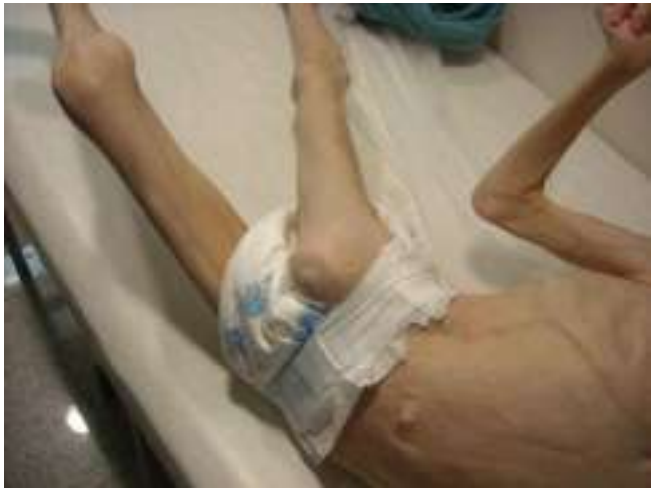
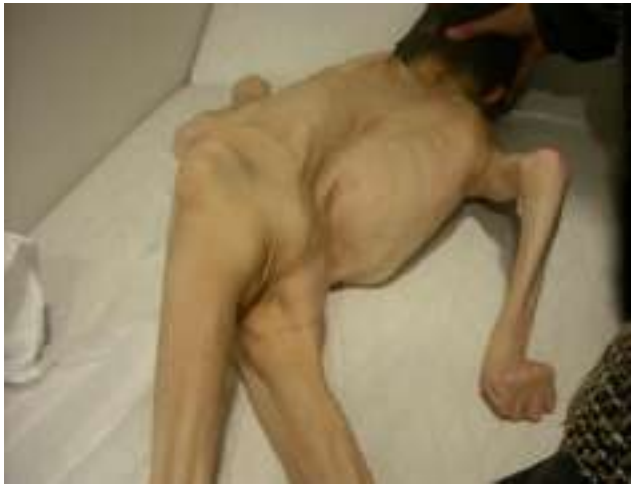
pelvic obliquity

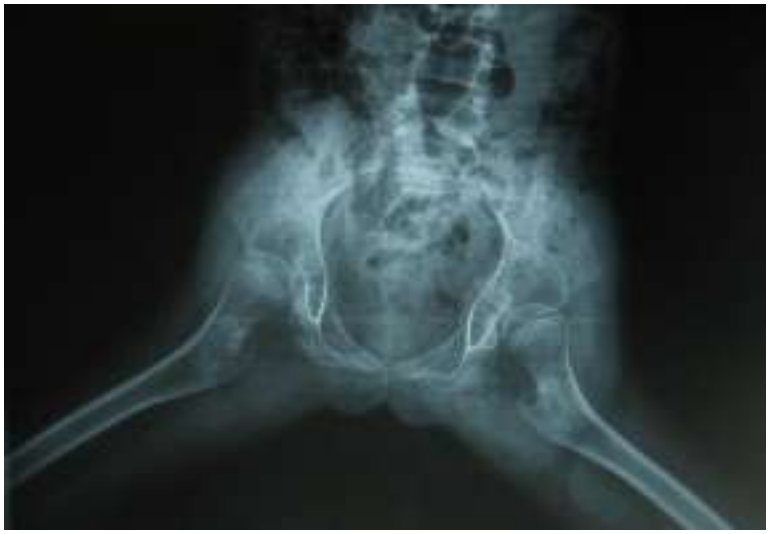




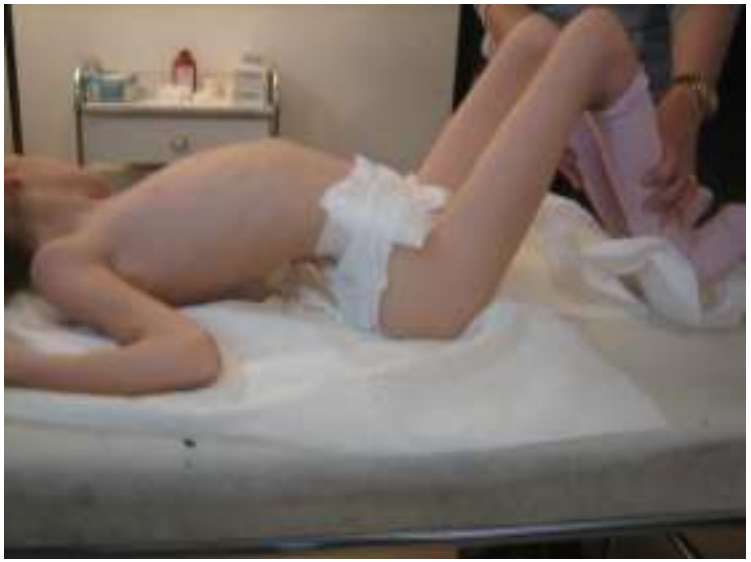
Spasticity in total involvement

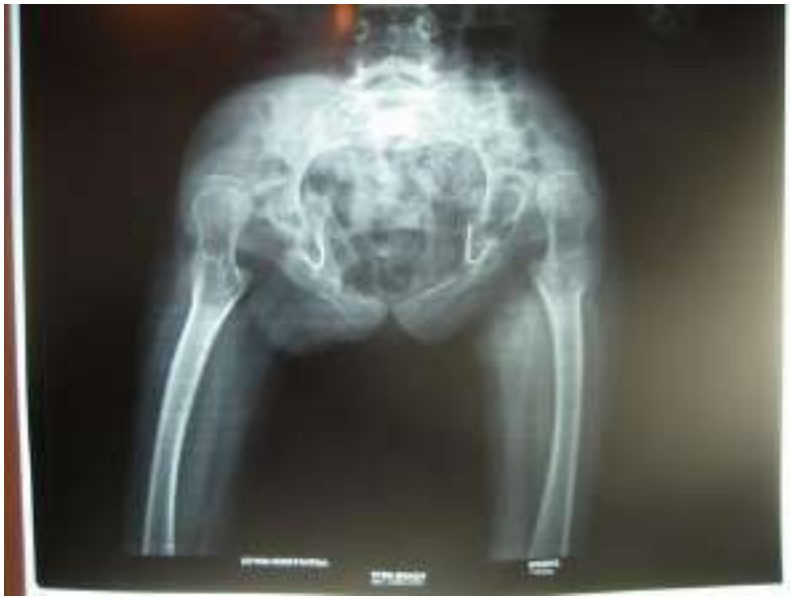






Hip dislocation - pelvic lordosis





Hip dislocation - pelvic lordosis

J Spine Surg. 2016 Dec; 2(4): 299–309.

The management of scoliosis in children with cerebral palsy: a review

Thomas Cloake and Adrian Gardner

- Children who suffer with cerebral palsy (CP) have a significant chance of developing scoliosis during their early years and adolescence. The behavior of this scoliosis is closely associated with the severity of the CP disability and unlike idiopathic scoliosis, it continues to progress beyond skeletal maturity. Conservative measures may slow the progression of the curve, however, surgery remains the only definitive management option. Advances in surgical technique over the last 50 years have provided methods to effectively treat the deformity while also reducing complication rates. The increased risk of surgical complications with these complex patients make decisions about treatment challenging, however with careful pre-operative optimization and post-operative care, surgery can offer a significant improvement in quality of life.

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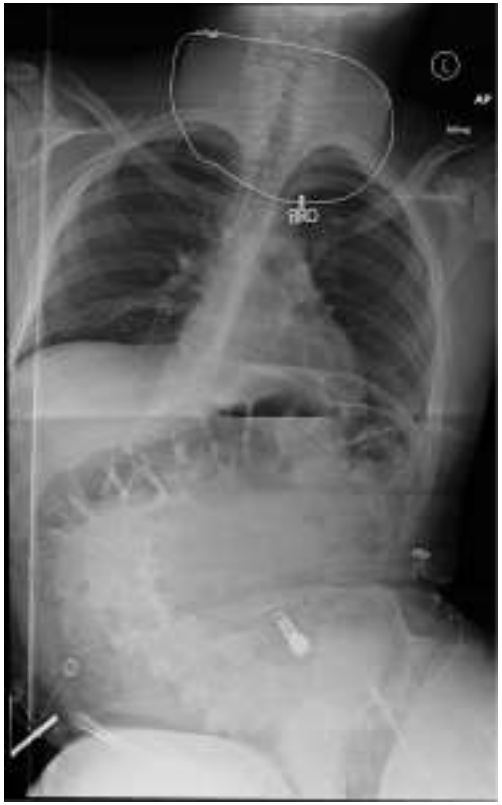
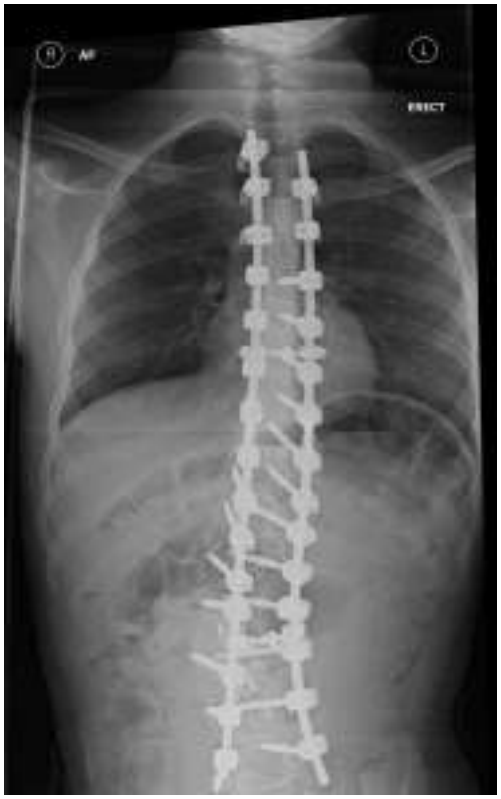
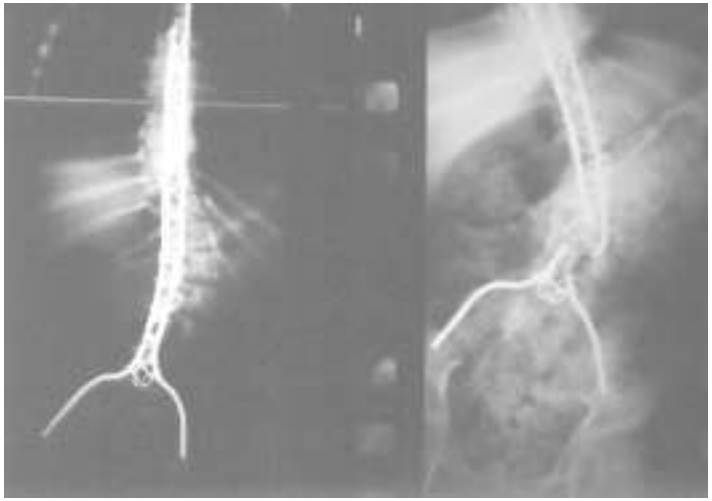
cerebral palsy





Surgical management of scoliosis in CP



Spasticity has tremendous strength, creating pseudoarthrosis

Indian J Orthop. 2010 Apr-Jun; 44(2): 148–158.

Development and treatment of spinal deformity in patients with cerebral palsy

Athanasios I Tsirikos, Consultant Orthopaedic and Spine Surgeon

- Spinal deformity in patients with severe neurological handicaps can affect their ability to sit and cause significant back pain or pain due to rib impingement against the elevated side of the pelvis on the concavity of the curvature.
- Surgical correction followed by spinal arthrodesis is indicated in patients with progressive deformities which interfere with their level of function and quality of life. Spinal deformity correction is **a major task** in children with multiple medical co-morbidities and can be associated with **a high risk of complications including death**

Indian J Orthop. 2010 Apr-Jun; 44(2): 148–158.

Development and treatment of spinal deformity in patients with cerebral palsy

Athanasios I Tsirikos, Consultant Orthopaedic and Spine Surgeon

- A **well-coordinated multidisciplinary approach** is required in the assessment and treatment of this group of patients with the aim to minimize the complication rate and secure a satisfactory surgical outcome
- Good knowledge of the surgical and instrumentation techniques, as well as the principles of management is needed **to achieve optimum correction of the deformity and balancing of the spine and pelvis**. Spinal fusion has a well-documented positive impact even in children with quadriplegia or total body involvement and is the only surgical procedure which has such a high satisfaction rate among parents and caregivers.

Dev Med Child Neurol. 2017 Jul;59(7):690-698.

Outcomes after scoliosis surgery for children with cerebral palsy: a systematic review.

Toovey R[1], Harvey A[1,2,3], Johnson M[4], Baker L[2], Williams K

- Limited high-quality evidence exists for outcomes after scoliosis surgery in children with CP, a procedure associated with a moderately high complication rate.
- The intervention appears indicated for deformity correction, but currently there is insufficient evidence to make recommendations for this surgery as a way to also improve functional outcomes, caregiver outcomes, and quality of life.

Acta Med Okayama. 2017 Apr;71(2):119-126.

Scoliosis in Patients with Severe Cerebral Palsy: Three Different Courses in Adolescents.

Oda Y[1], Takigawa T, Sugimoto Y, Tanaka M, Akazawa H, Ozaki T.

- Thirty four severe CP patients presented with scoliosis and were divided into 3 groups based on their clinical courses: severe, moderate and mild.
- The mean Cobb angles at the final follow-up were **129°,** 53°, and 13° in the severe, moderate, and mild groups, respectively

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Neuromuscular diseases

- Spinal motor neuron

SMA

- Distal nerve motor and sensory neuropathies

HMSNs

- Muscle progressive disease

muscular dystrophies , arthrogryposis

Primary muscle disease
Progressive muscular dystrophies

- Duchenne muscular dystrophy DMD
- Becker muscular dystrophy
- Emery Dreifuss muscular dystrophy
- Facio scapulo humeral dystrophy
- Limb Girdle muscular dystrophy

Duchenne muscular dystrophy
Ambulation period

- Orthopaedic management in Duchennne MD

Η ορθοπεδική προσέγγιση των παιδιών με μυϊκή δυστροφία Duchenne

Ν. ΛΑΛΙΩΤΗΣ

University Orthopaedic Dept. Alder Hey Children's Hospital, Liverpool England

ABSTRACT: «The orthopaedic management of Duchenne muscular dystrophy».

N. LALIOTIS

Duchenne muscular dystrophy, an X-linked inherited disorder, is characterised with progressive degeneration and weakness of the skeletal muscle. The disease progresses with characteristic postural and walking changes, contractures of muscles and finally children cannot compensate any more. Walking ability ceases, after which children become weel-chair bound. Gait analysis can record accurately the progression of the disease, mainly with the increase in double stance time. Physiotherapy, calipers and early surgical release of tight muscles, are used in order to prolong the walking period. When the child is no more able to stand, scoliosis is the major orthopaedic problem. Early surgical stabilisation of the spine, when respiratory function is still acceptable, can improve the quality of life of these children. The orthopaedic surgeon has an important role in the group that treat these children and in the early detection of the disease.

Key words: Duchenne muscular dystrophy, treatment, gait analysis.

ΠΕΡΙΛΗΨΗ: Η μυϊκή δυστροφία Duchenne χαρακτηρίζεται από εξελισσόμενη αδυναμία βάδισης και στή-

ριξης του παιδιού, που καταλήγει σε αναπηρική καρέκλα στην δεύτερη δεκαετία της ζωής. Οι μεταβολές της κινητικής δραστηριότητας αντανακλούν τα στάδια παράλυσης των διαφόρων μυϊκών ομάδων. Η ανάλυση βάδισης καταγράφει αντικειμενικά την εξέλιξη της νόσου, μετράντας κυρίως την σταδιακή αύξηση του χρόνου διπλής στήριξης. Η φυσιοθεραπεία, η εφαρμογή ναρθήκων αλλά και η πρώιμη διατομή των ραιδεμένων μυών, αποσκοπούν στην παράταση του χρόνου της αυτόνομης στήριξης.

Όταν το παιδί χάσει την ικανότητα όρθιας στήριξης, η σκολίωση είναι το μείζον ορθοπεδικό πρόβλημα. Η πρώιμη χειρουργική αντιμετώπιση, όταν υπάρχει ακόμη επαρκής ζωτική χωρητικότητα των πνευμόνων, προσφέρει παύση και παράταση στην ζωή του ασθενούς.

Ο ορθοπεδικός έχει σημαντικό ρόλο στην ομάδα που αντιμετωπίζει τα παιδιά με μυϊκή δυστροφία, αλλά βοηθά σημαντικά και στην έγκαιρη διάγνωση της νόσου.

Λέξεις Κλειδιά: Μυϊκή δυστροφία Duchenne, θεραπεία, ανάλυση βάδισης.

Εισαγωγή

Οι μυϊκές δυστροφίες αποτελούν ένα σύνολο κληρονομικών, μη φλεγμονωδών, παθήσεων, που χαρακτηρίζονται από εξελισσόμενη αδυνα-

Duchenne muscular dystrophy scoliosis

- Appears with the loss of the standing position
- Early deterioration
- Paralytic C curve
- Inability to sit properly

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Duchenne muscular dystrophy scoliosis

- Early surgical intervention because of the respiratory failure
- Improvement of life expectancy and quality of daily life

Duchenne muscular dystrophy scoliosis

- Requires long fusion to pelvis
- Technically demanding
- Blood loss
- Impairment of FVC ?

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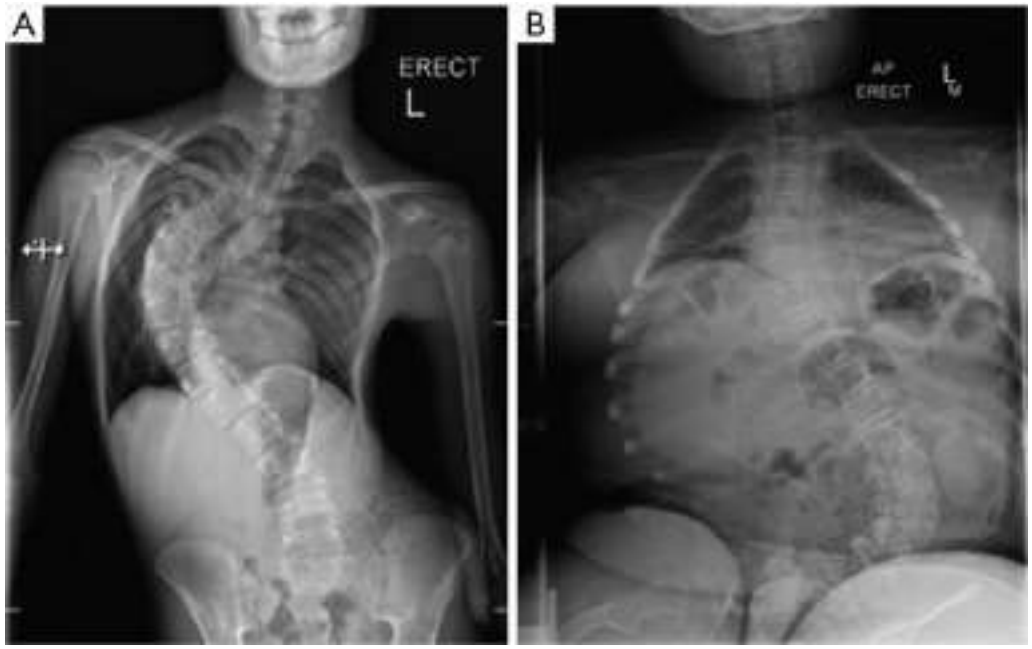
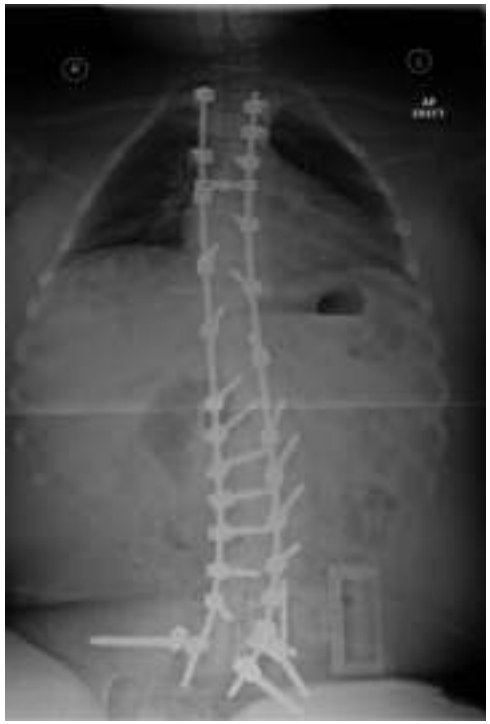
Duchenne muscular dystrophy





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Neuromuscul Disord. 2013 Aug;23(8):611-7.

Scoliosis in Duchenne muscular dystrophy (DMD).

Hsu JD[1], Quinlivan R.

.

- Scoliosis is a frequent complication in the non-ambulant patient with Duchenne muscular dystrophy (DMD). Weakness of the paraspinal muscles leads to trunk and body positional changes facilitating the development of a progressive collapsing scoliosis which inevitably interferes with comfortable sitting and may exacerbate deteriorating respiratory function. The recommended international standard of care for management of DMD **includes strategies to prolong ambulation which may delay the onset of scoliosis.** In the non-ambulant child there should be regular monitoring for scoliosis and, when present**, surgical treatment should undertaken at an early stage**. Careful multi-disciplinary pre-operative assessment and peri-operative care are essential.

J Pediatr Rehabil Med. 2016;9(1):23-9.

Management of scoliosis in patients with Duchenne muscular dystrophy and spinal muscular atrophy: A literature review.

Garg S.

.

- **Scoliosis occurs in nearly all non-ambulatory children with spinal muscular atrophy (SMA) and Duchenne muscular dystrophy (DMD).** Non-operative treatments have not been shown to be effective at preventing progression of scoliosis. Progressive scoliosis can impact the ability of patients to sit comfortably, be cosmetically unappealing, and in severe cases exacerbate pulmonary disease. The main goal of operative treatment is to improve sitting balance and prevent progression of scoliosis. Complication rates are high and there is little data on effect of operative treatment on quality of life in children with SMA and DMD. Comprehensive multi-disciplinary pre-operative evaluations are vital to reduce the risks of operative treatment

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Cochrane Database Syst Rev. 2015 Oct 1;(10).

Surgery for scoliosis in Duchenne muscular dystrophy.

Cheuk DK[1], Wong V, Wraige E, Baxter P, Cole A.

- Since no randomized controlled clinical trials were available to evaluate the effectiveness of scoliosis surgery in patients with DMD, we can **make no good evidence-based conclusion to guide clinical practice.** Patients with scoliosis should be *informed as to the uncertainty of benefits and potential risks of surgery for scoliosis*. Randomized controlled trials are needed to investigate the effectiveness of scoliosis surgery, in terms of quality of life, functional status, respiratory function, and life expectancy

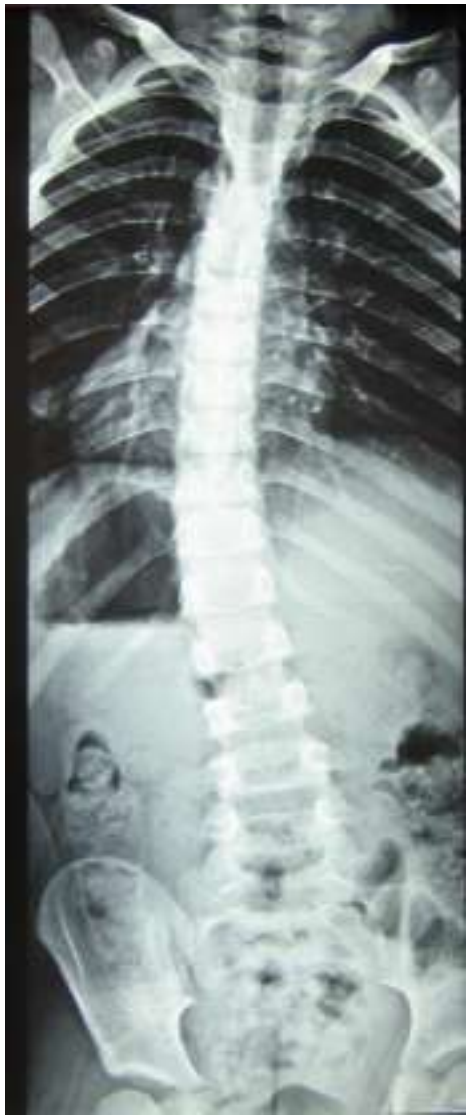
Pediatrics. 2018 Oct;142(Suppl 2):S82-S89.

Orthopedic and Surgical Management of the Patient With Duchenne Muscular Dystrophy.

Apkon SD[1], Alman B[2], Birnkrant DJ[3], Fitch R[4], Lark R[4], Mackenzie W[5], Weidner N[6], Sussman M[7].

- In this review, we target pediatricians, neurologists, orthopedic surgeons, rehabilitation physicians, anesthesiologists, and other individuals involved in the management of patients with DMD by providing specific recommendations to guide clinical practice related to orthopedic issues and surgical management in this setting

Early brace in scoliosis Duchenne



Electric standing frames

Spinal muscular atrophy
Functional classification

- Patients never sit independently
- Patients with head control, but cannot stand independently
- Patients that stand by themselves and have limited walking ability with assistance
- Patients that can walk alone but no run or climb stairs

Evans, Drennan, Russman S. Functional classification and orthopaedic management of SMA J Bone Joint Surg Br 1981

arthrogryposis

- Multiple joint involvement
- Fixed joints
- Skin and muscle involvement
- Smooth skin, absent skin creases
- NON progressive lesion

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arthrogryposis

ΕΕΧΟΤ
Τόμος 57, (4):199-206, 2006

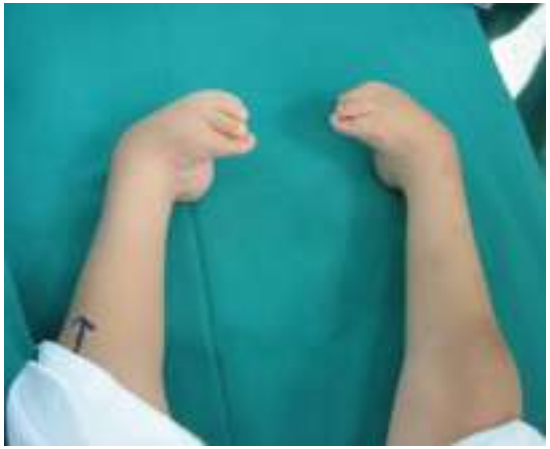
Αντιμετώπιση παιδιών με συγγενή αρθρογρύπωση. Προσβολή στα κάτω άκρα και κινητική εξέλιξη των παιδιών

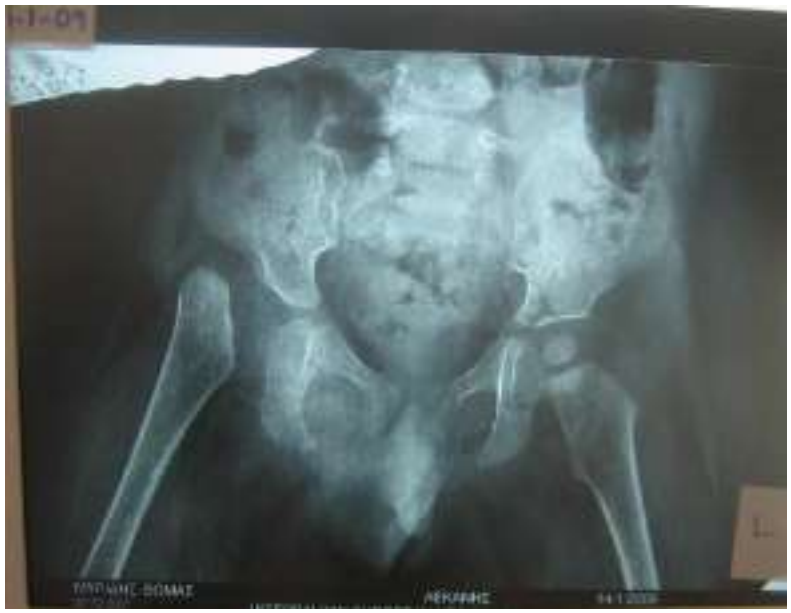
ΛΑΛΙΩΤΗΣ ΝΙΚΟΛΑΟΣ, ΜΥΛΩΝΑΣ ΧΡΗΣΤΟΣ, ΚΟΥΚΟΥΜΠΗΣ ΘΕΟΔΟΣΙΟΣ, ΟΙΚΟΝΟΜΙΔΗΣ ΓΕΩΡΓΙΟΣ

ΕΛΕΠΑΠ Θεσσαλονίκης, Γεν. Κλινική "Άγιος Λουκάς"

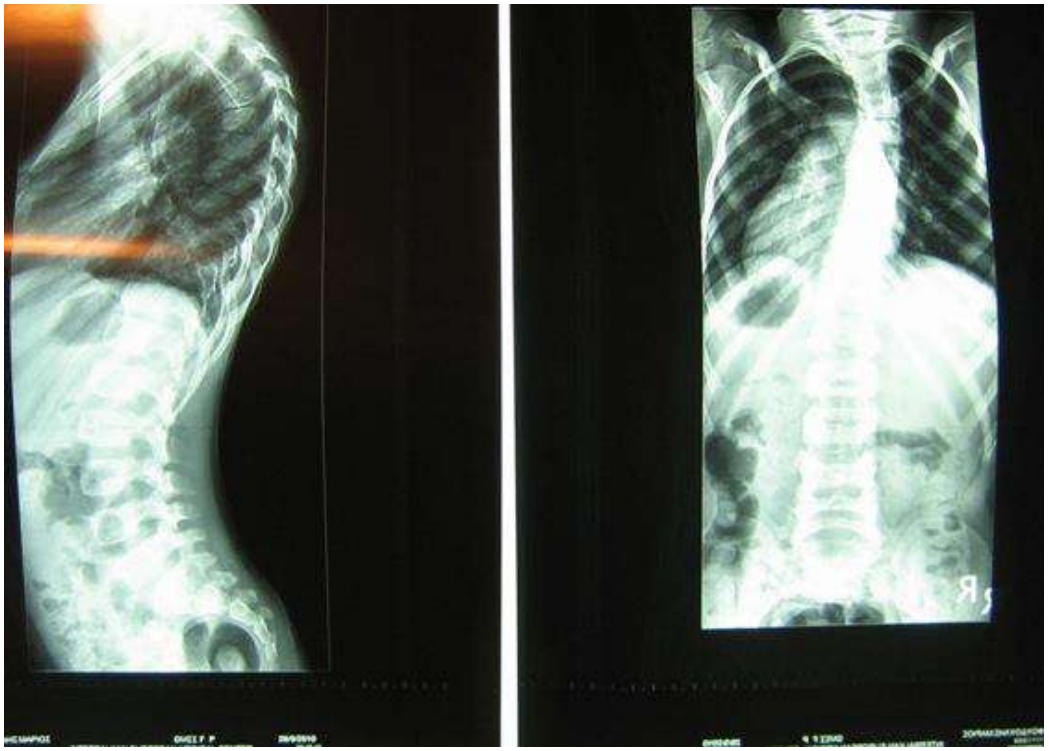
Arthrogryposis upper limb

Myrid thom intubation in arthrogryposis





Arthrogryposis scoliosis



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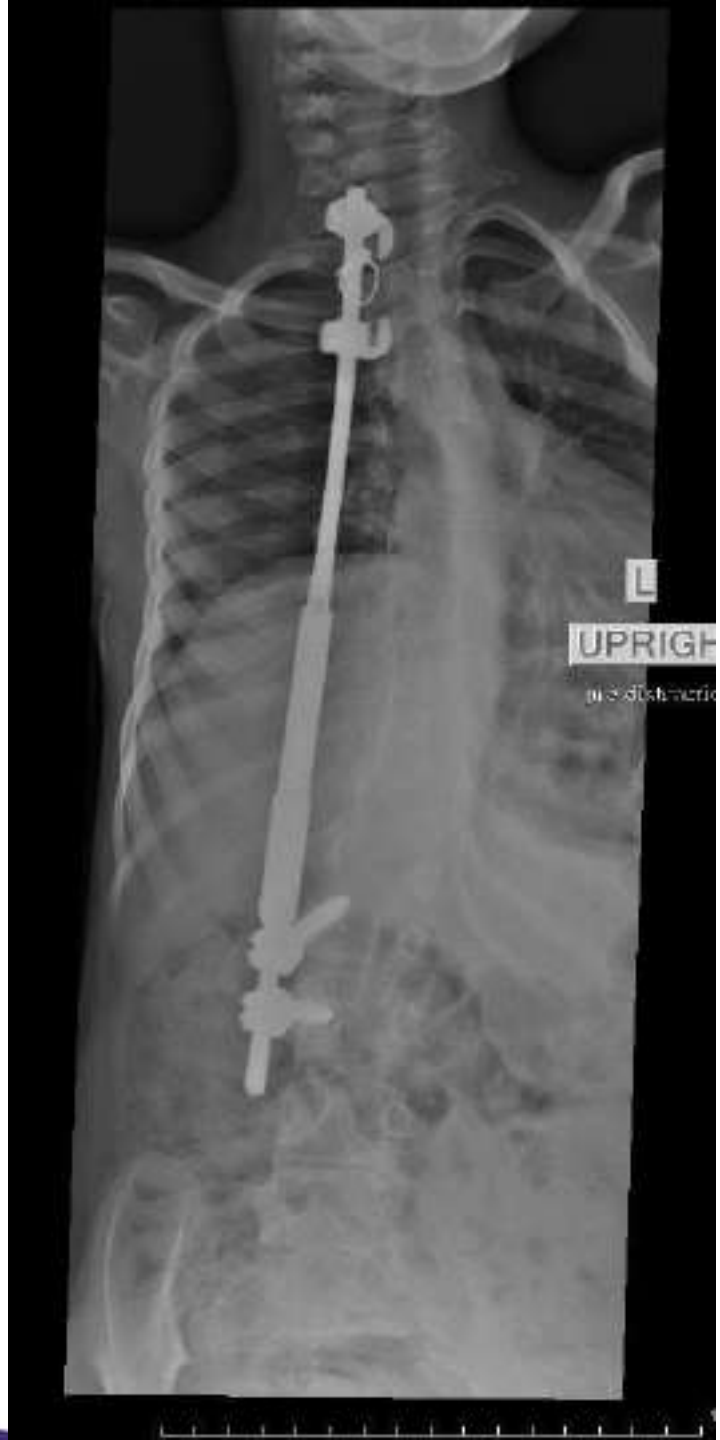
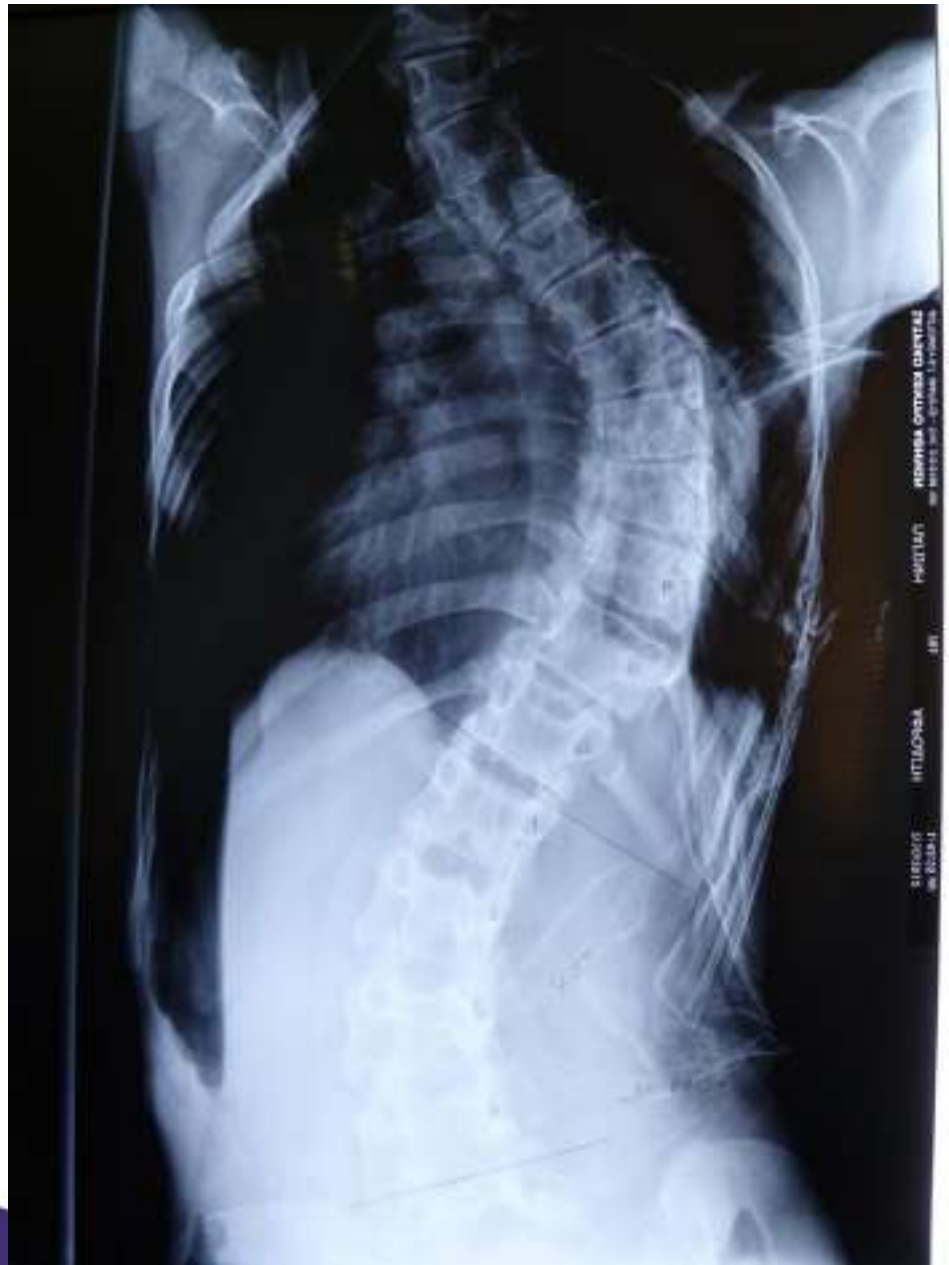


Diploma of the
Hellenic Spine Society
(equivalent to the Eurospine Diploma)

Hellenic
Spine
Society



Diploma of the
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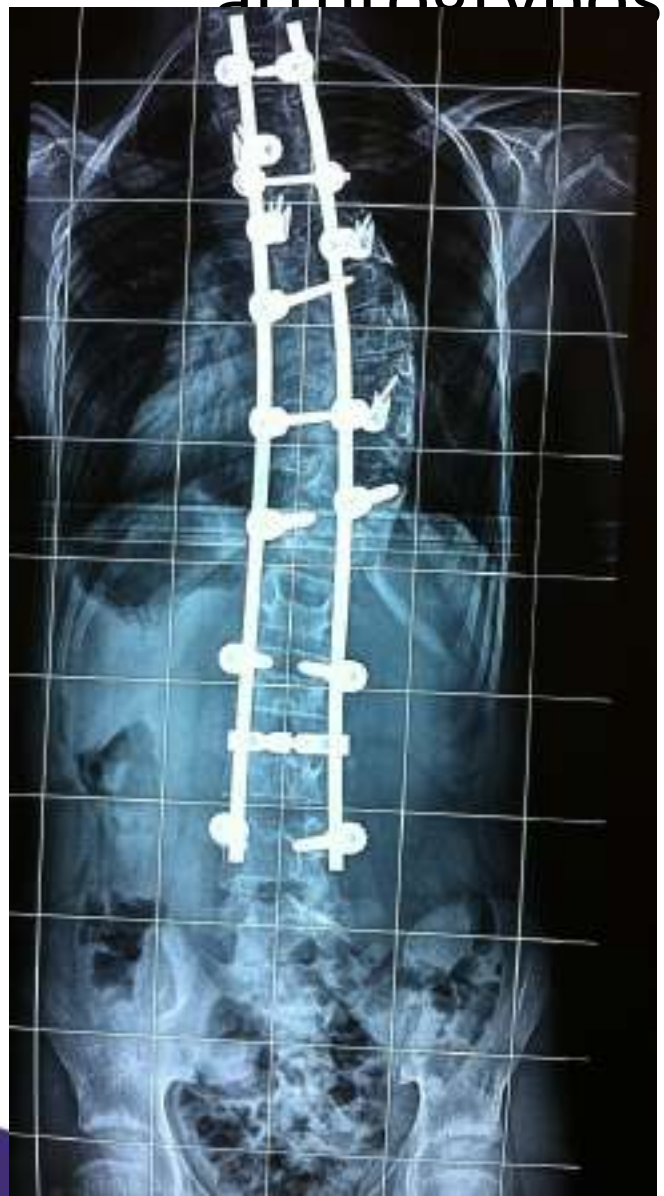

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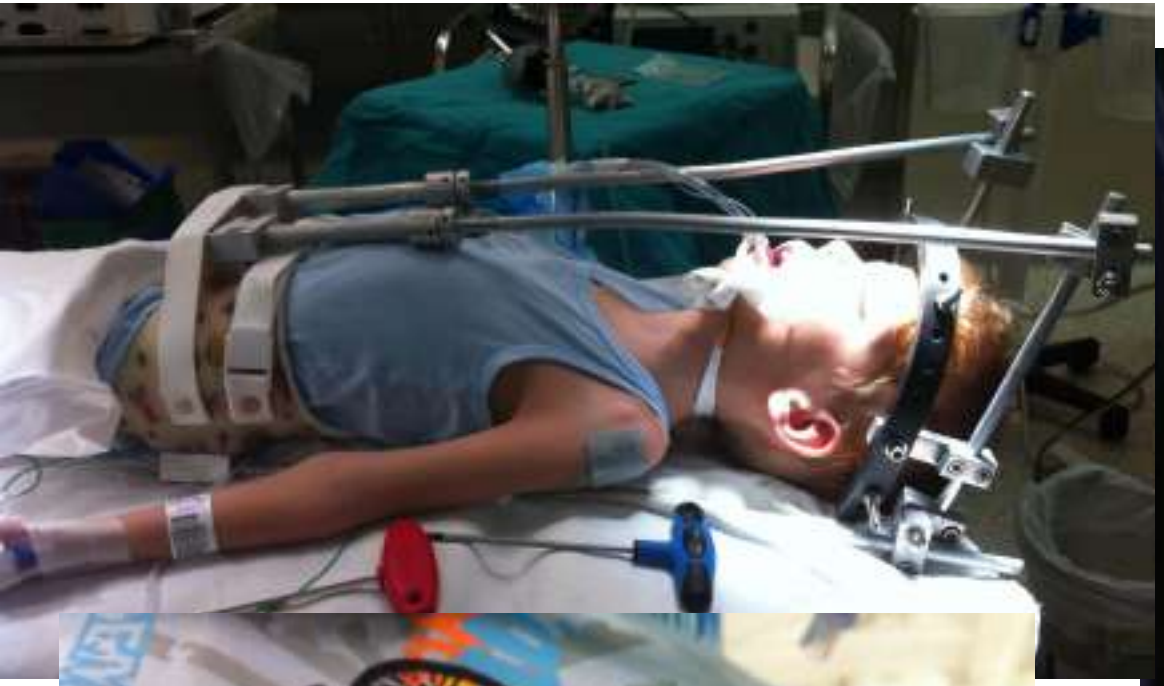

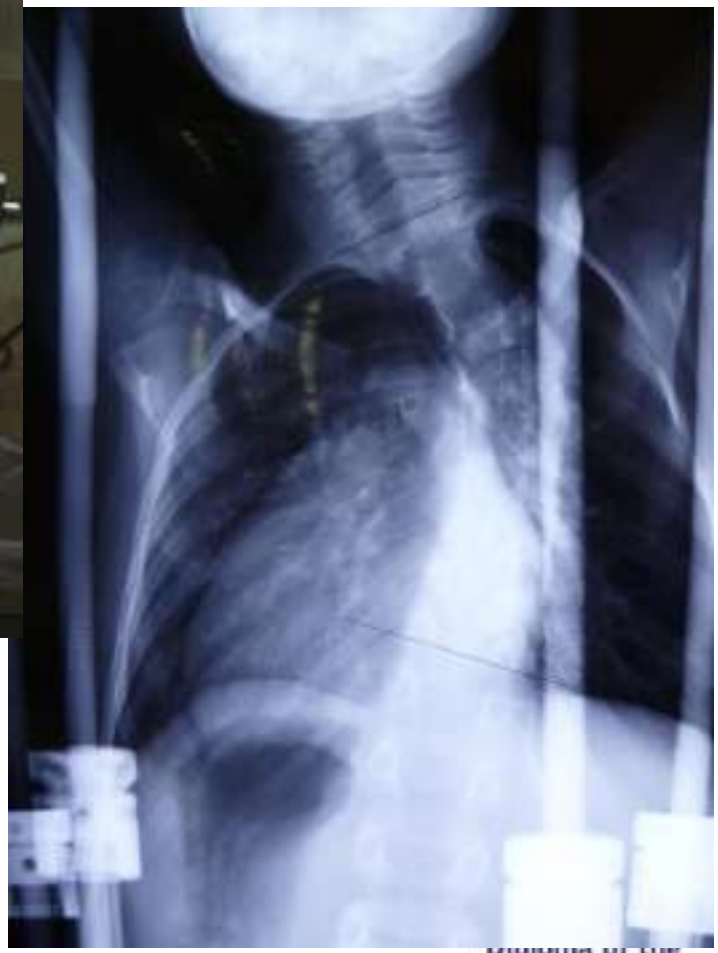

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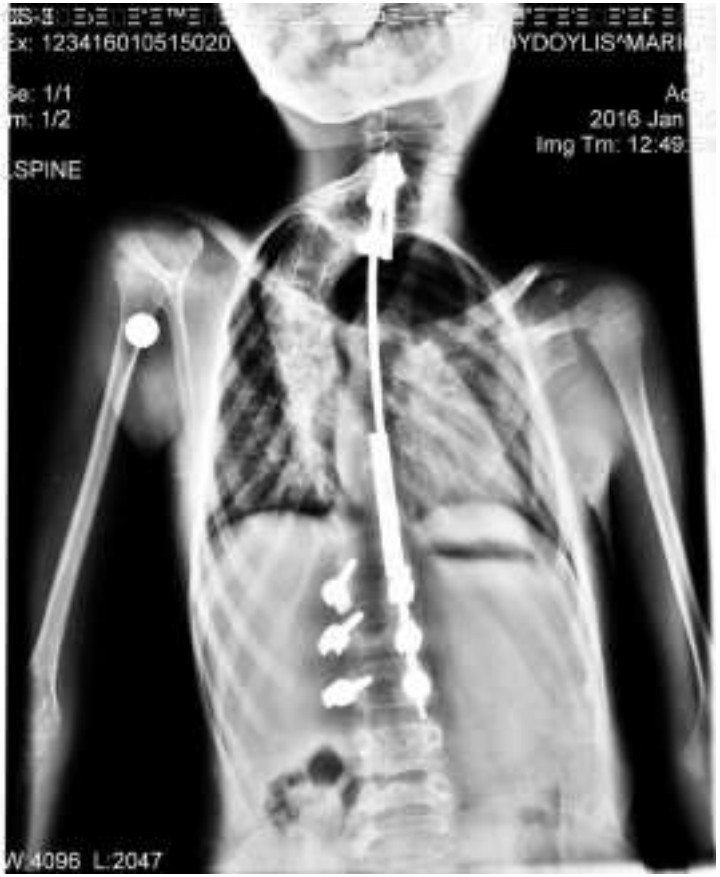

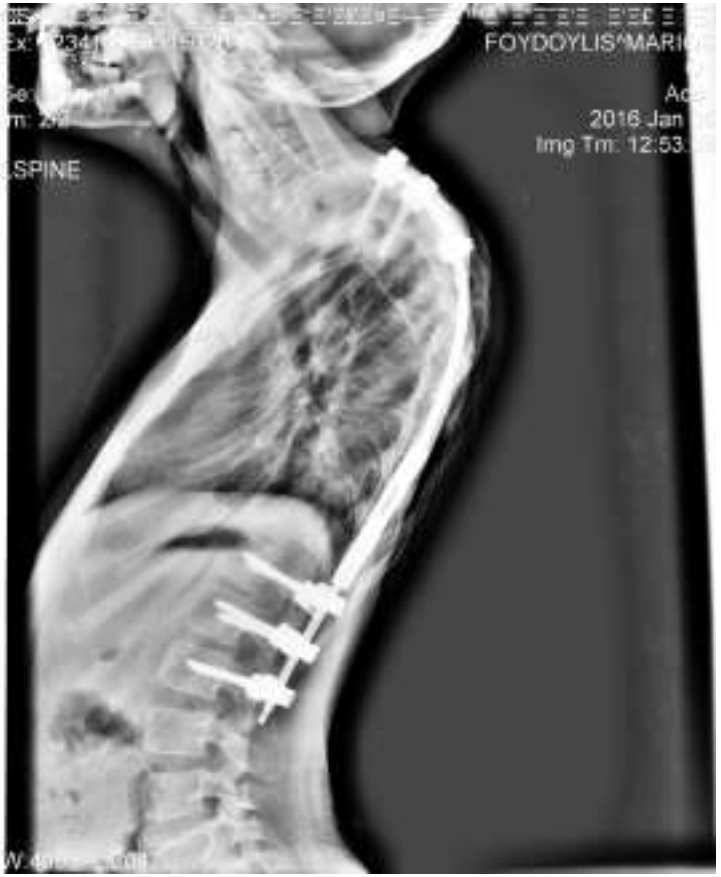

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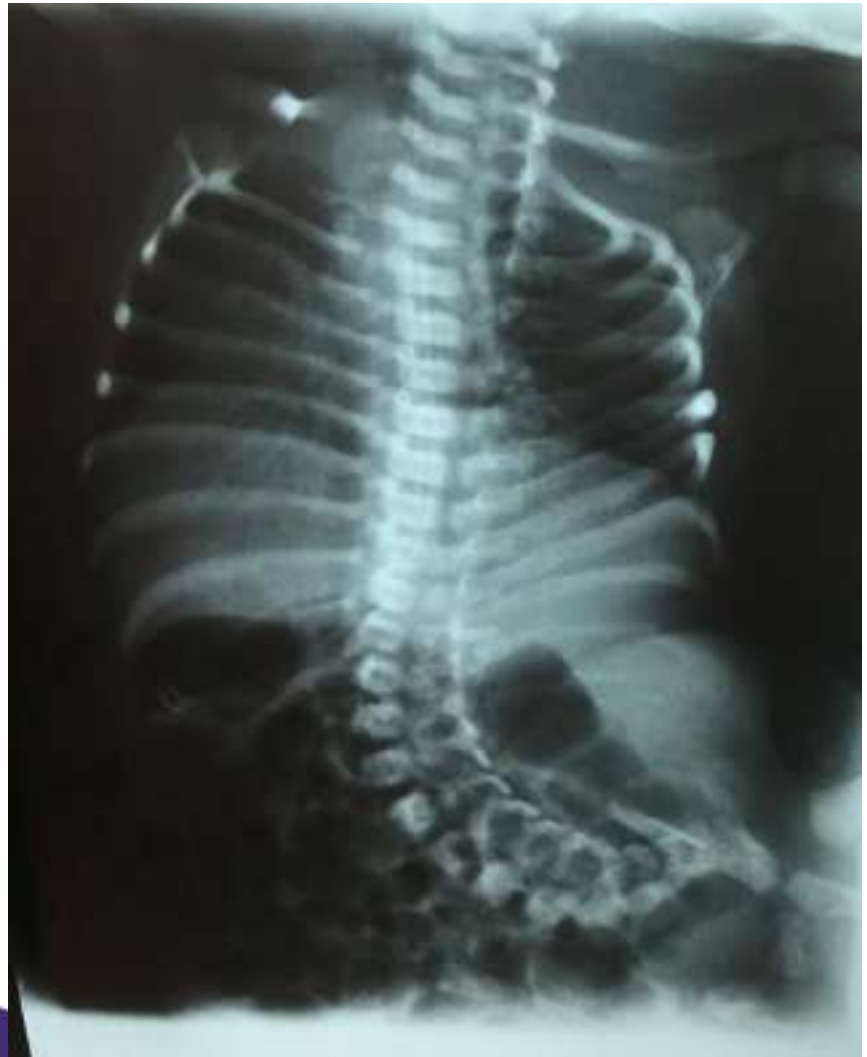

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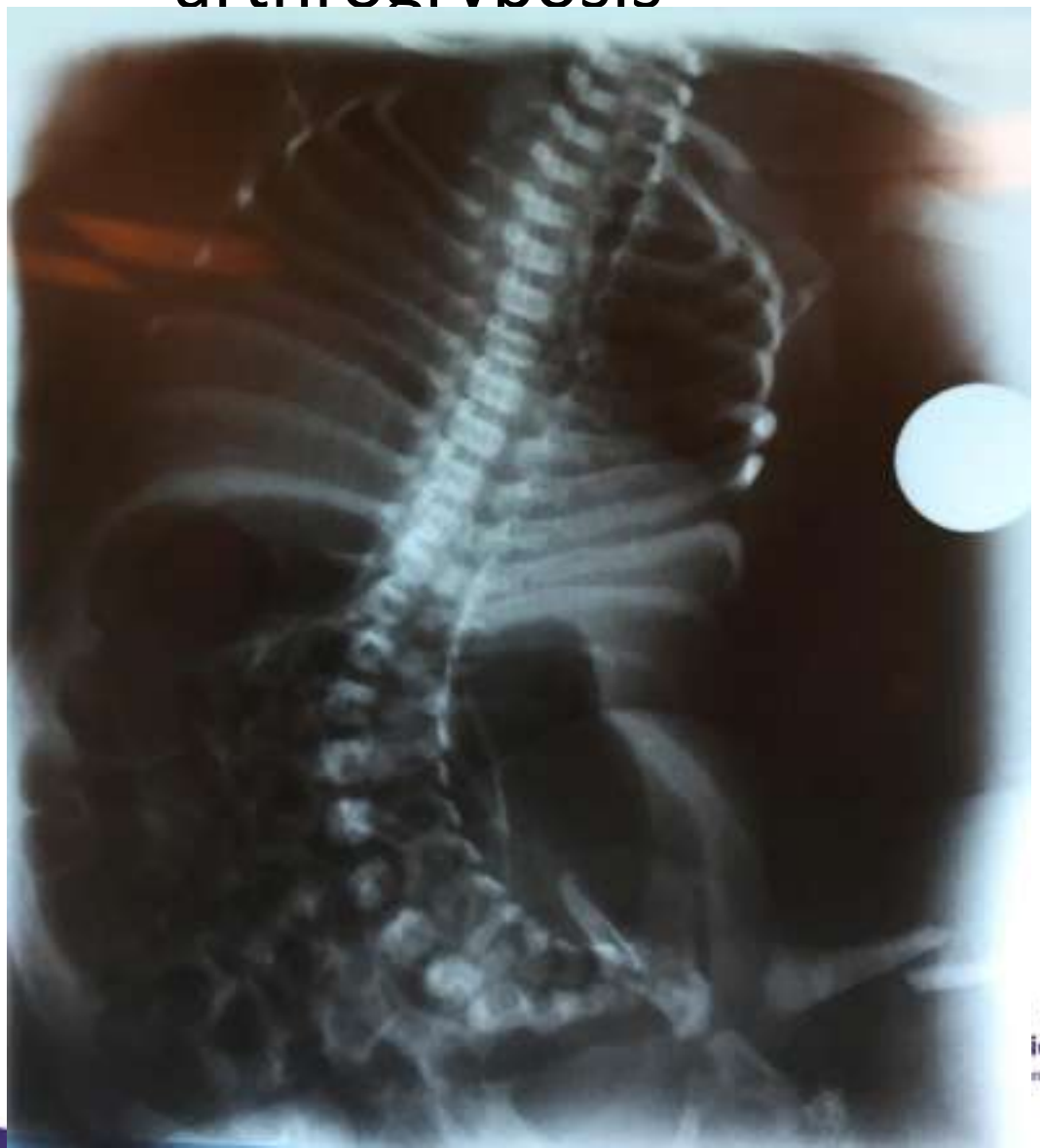

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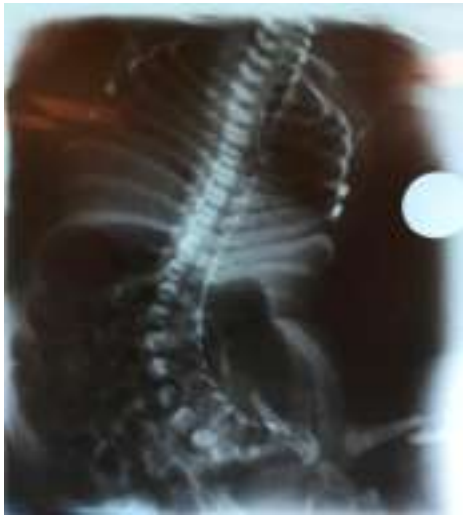

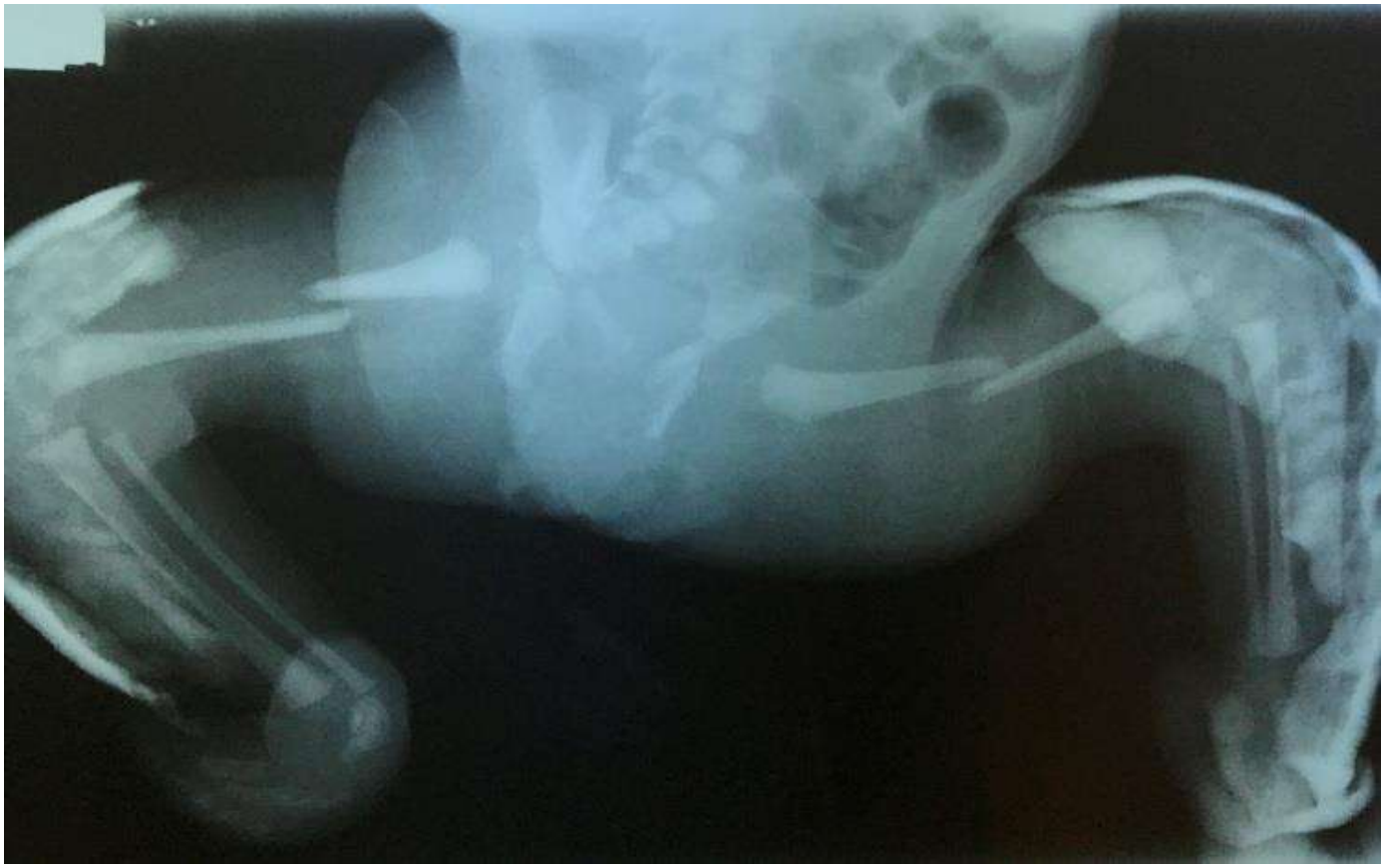

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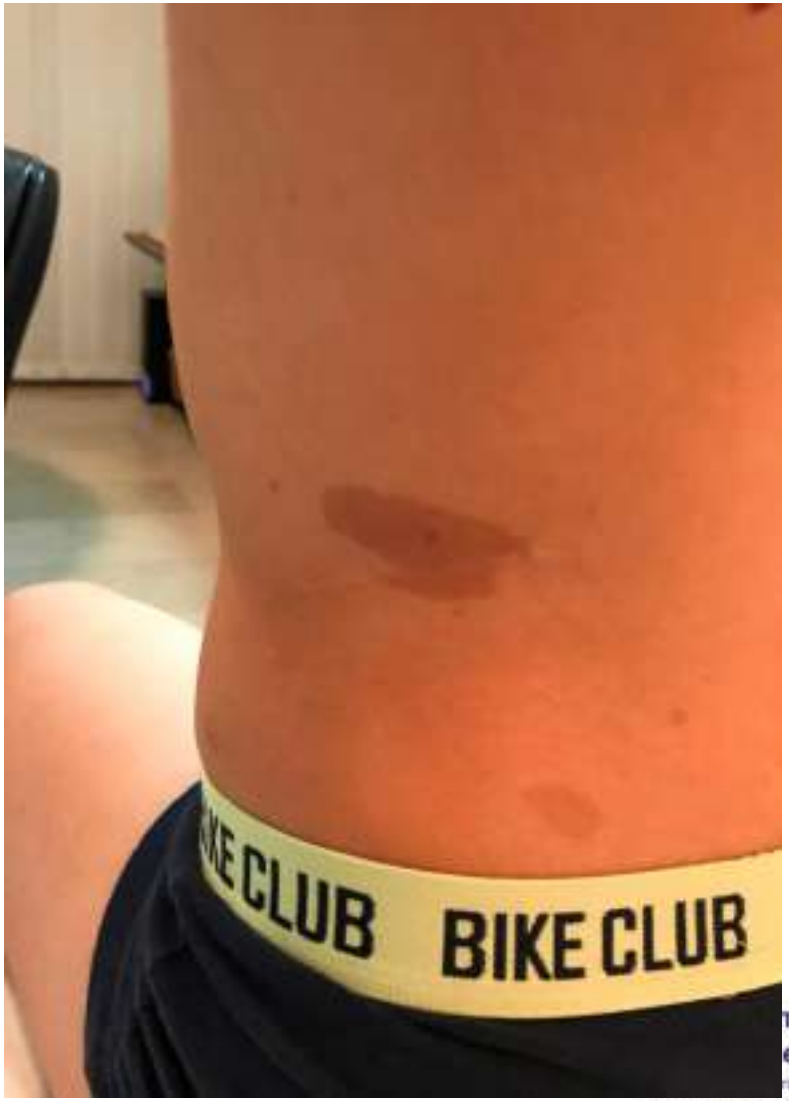

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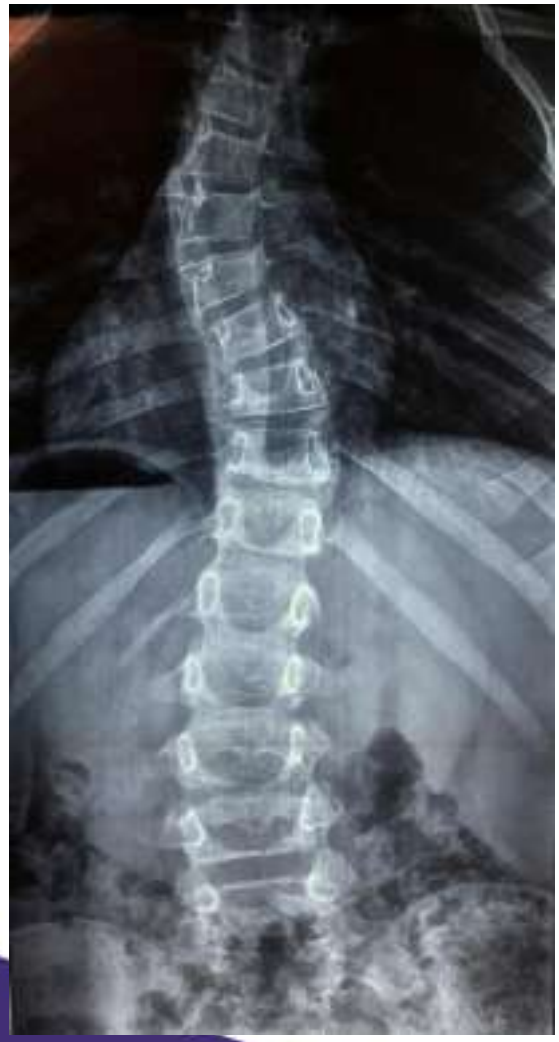
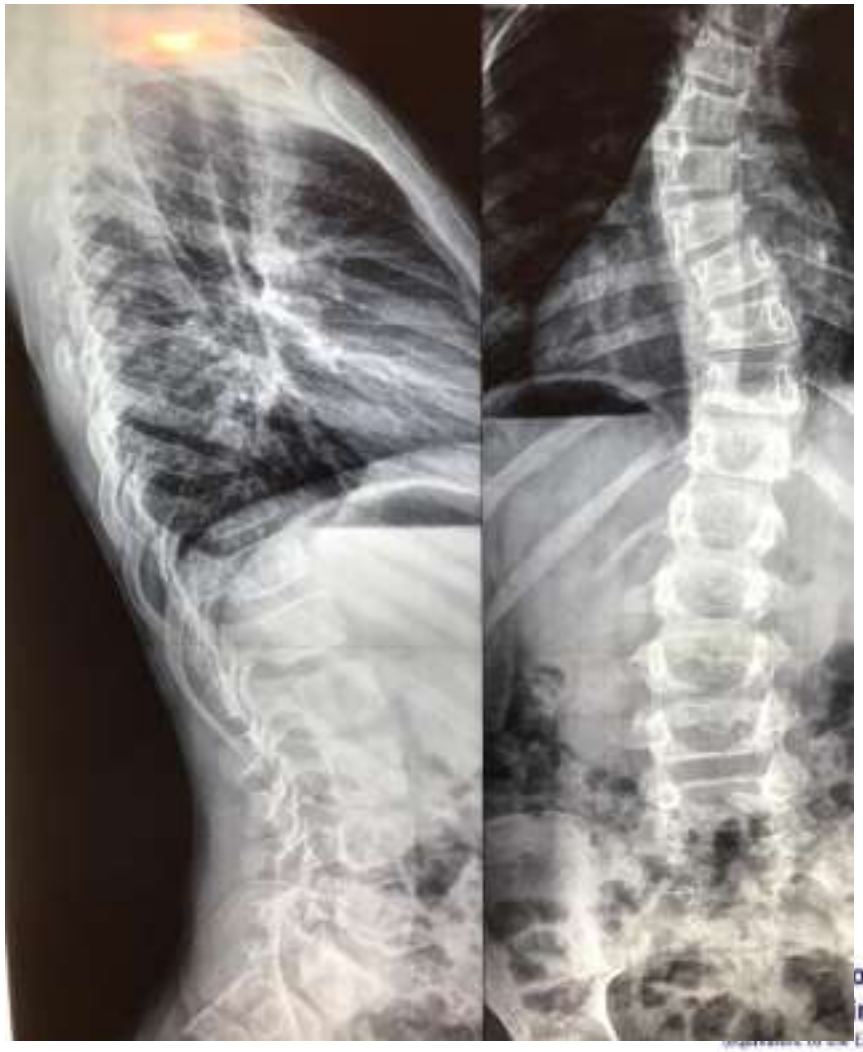
neurofibromatosis





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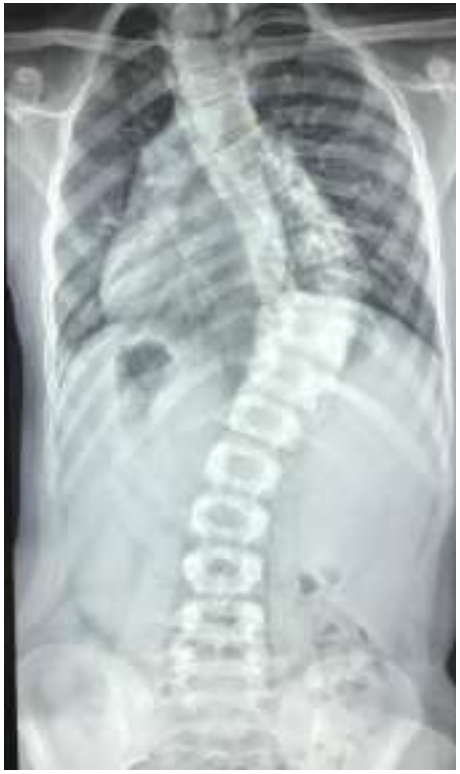
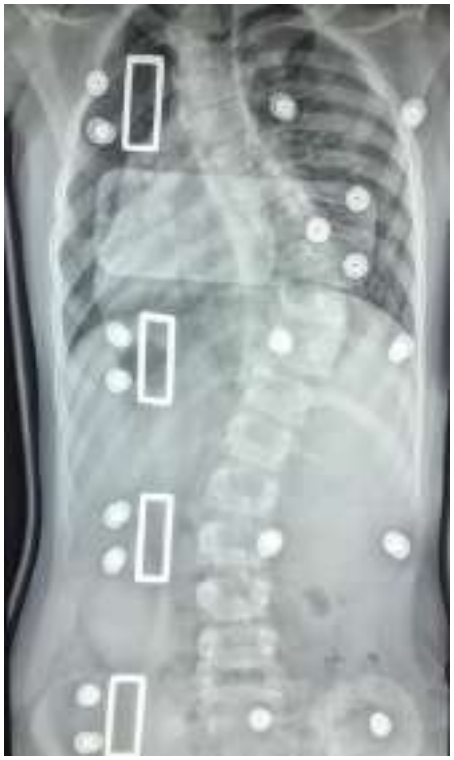
Neurofibromatosis dystrophic scoliosis





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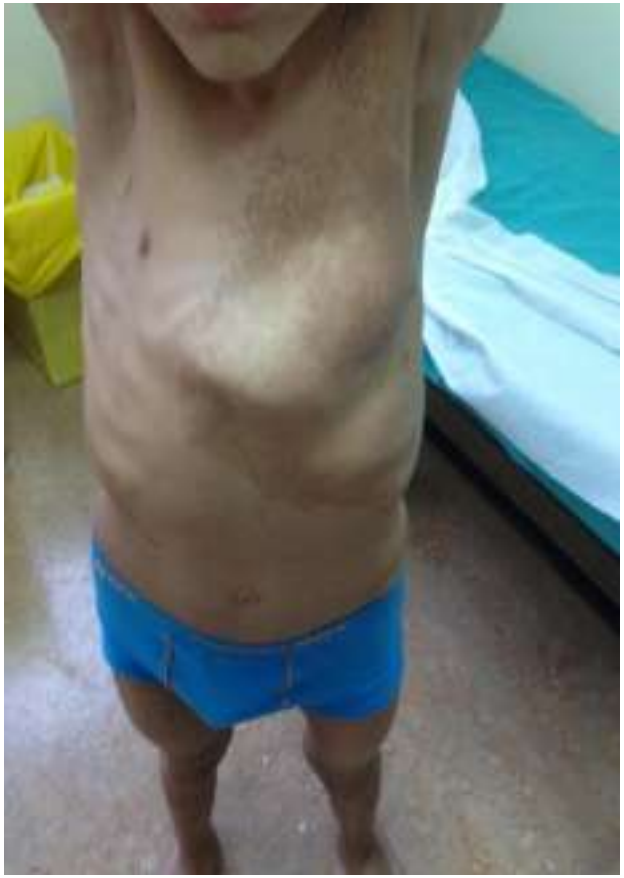
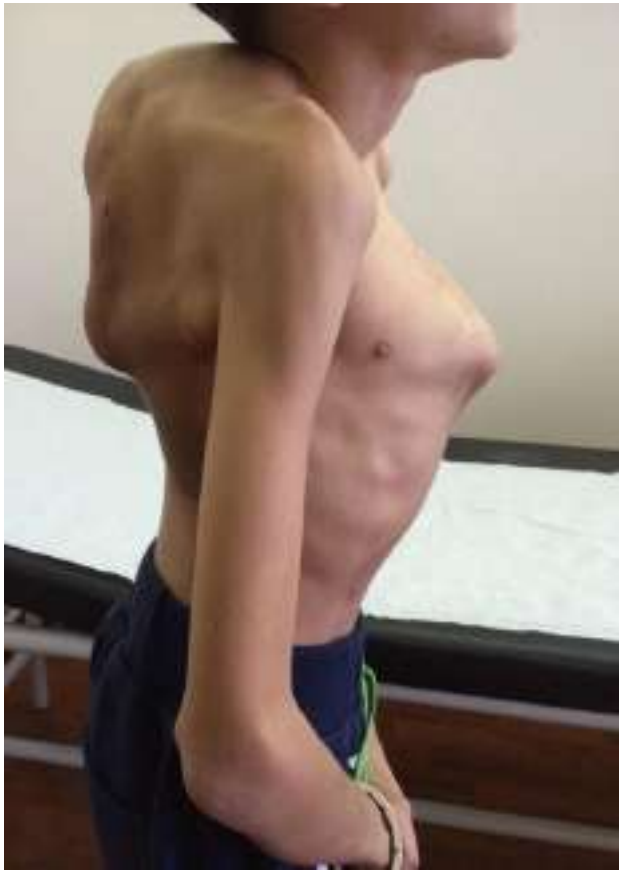
Neurofibromatosis fibromas





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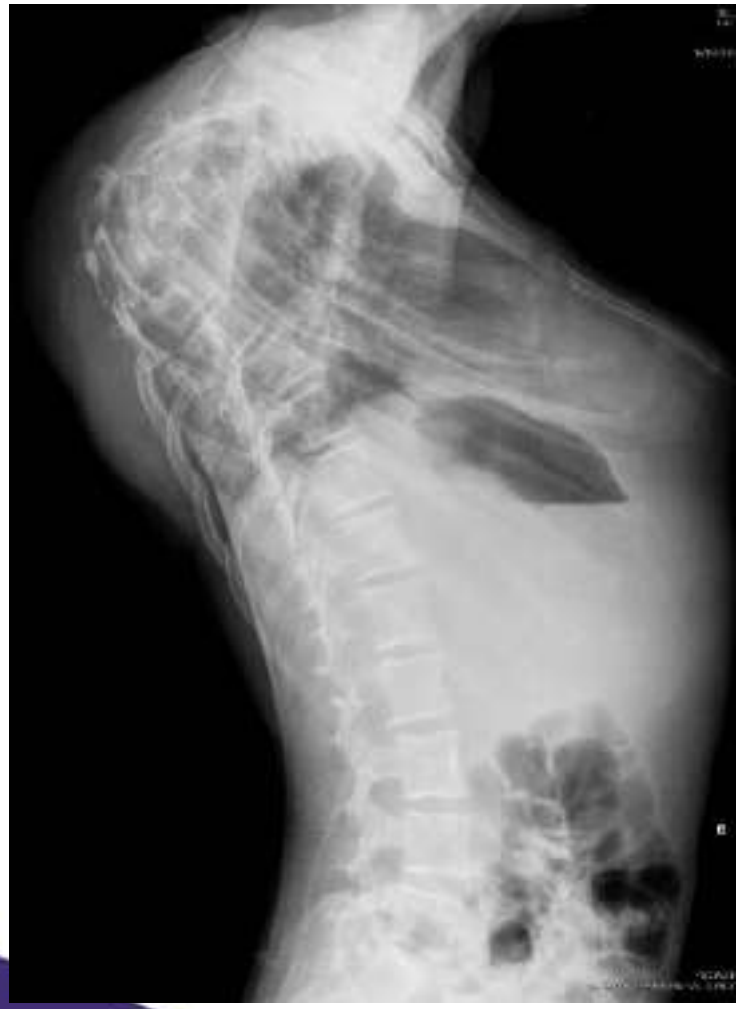
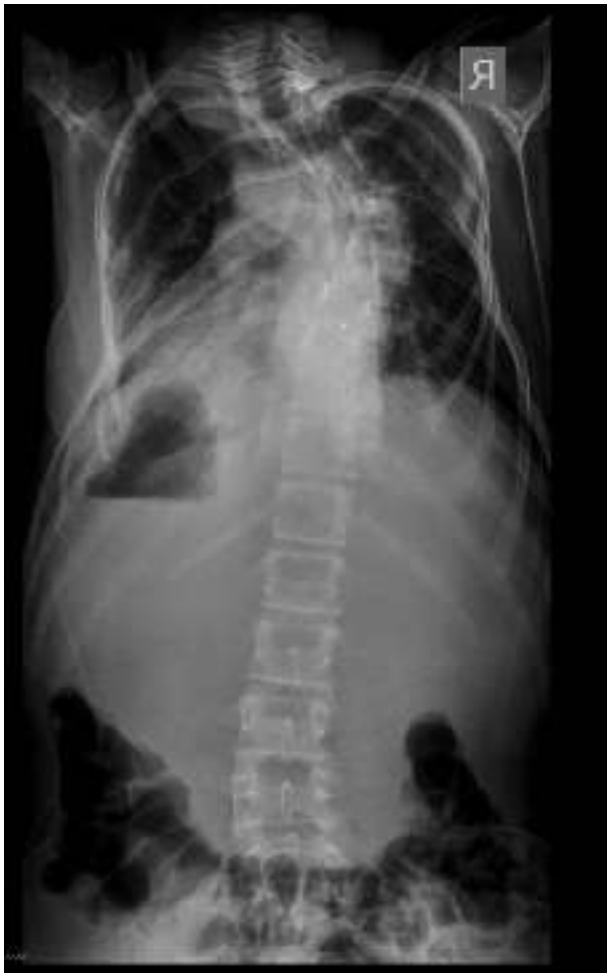
Neurofibromatosis fibromas





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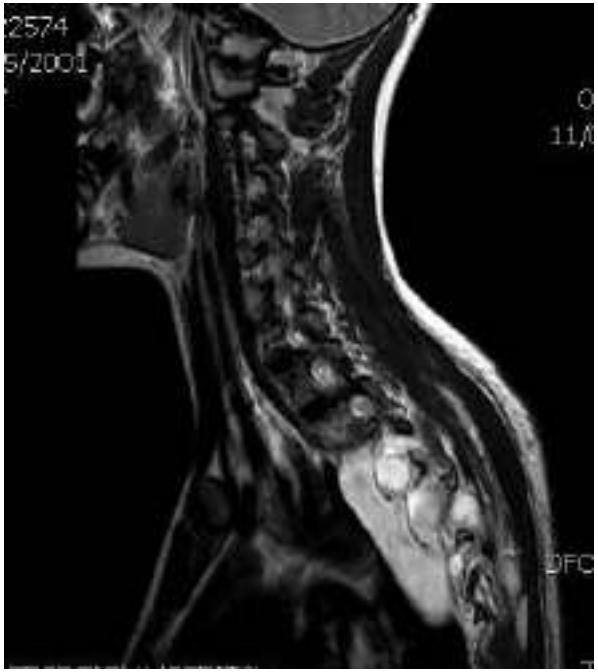
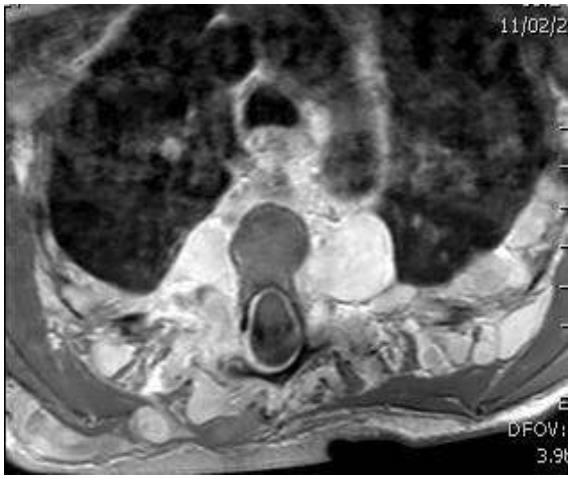
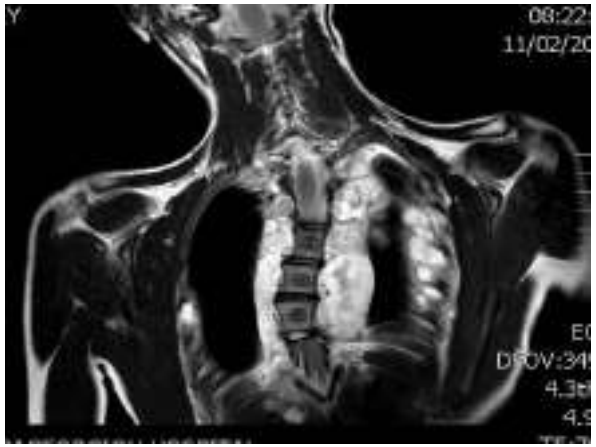
Neurofibromatosis fibromas







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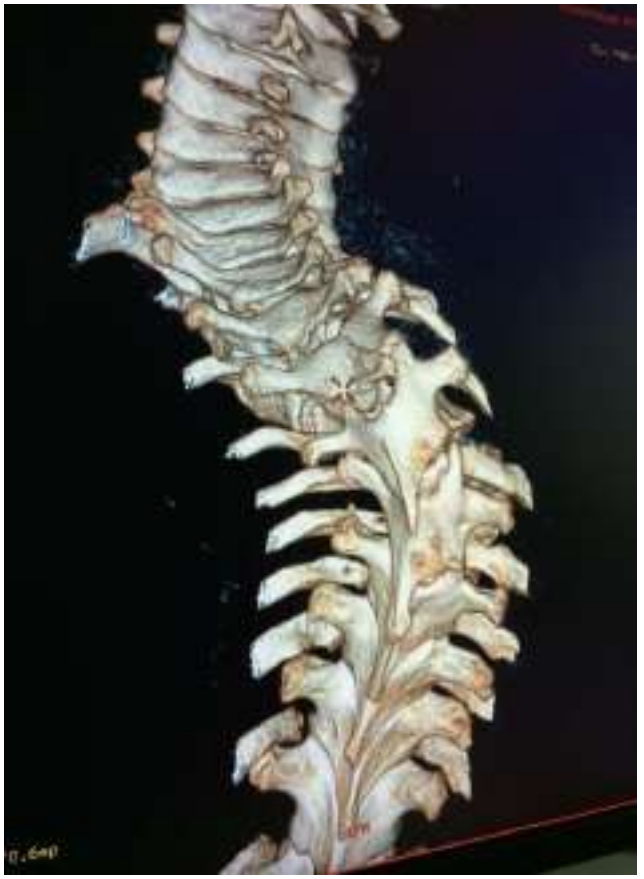
Neurofibromatosis fibromas



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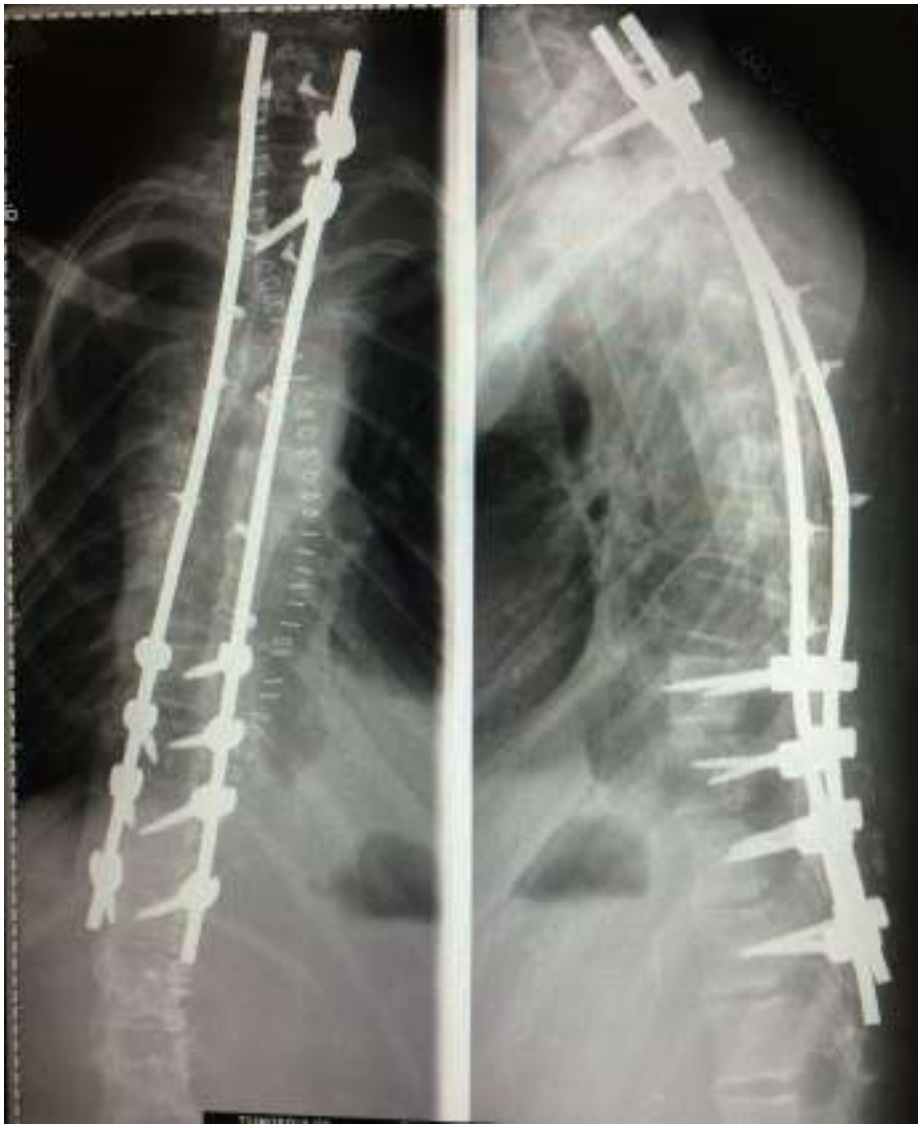

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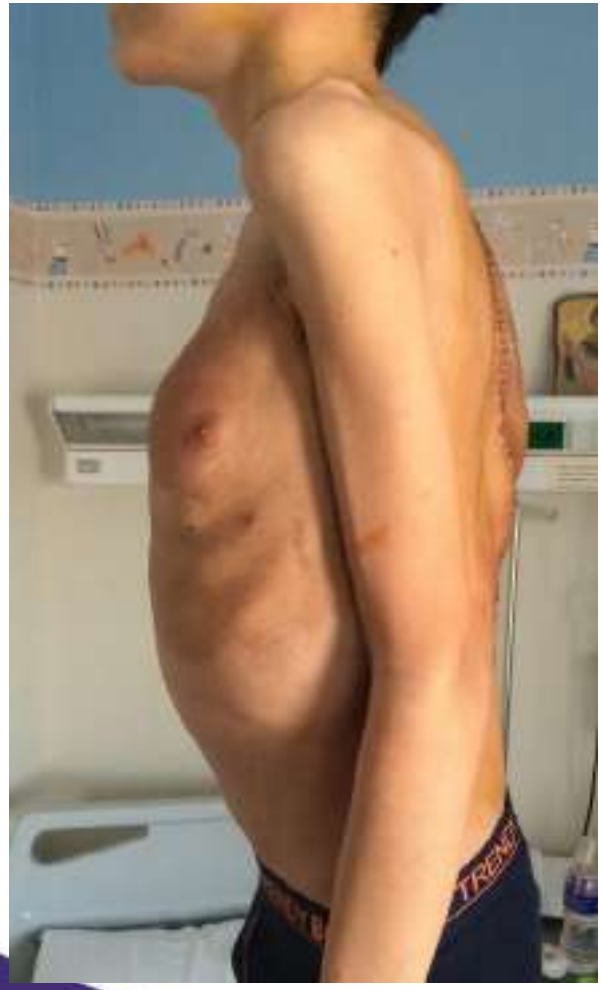

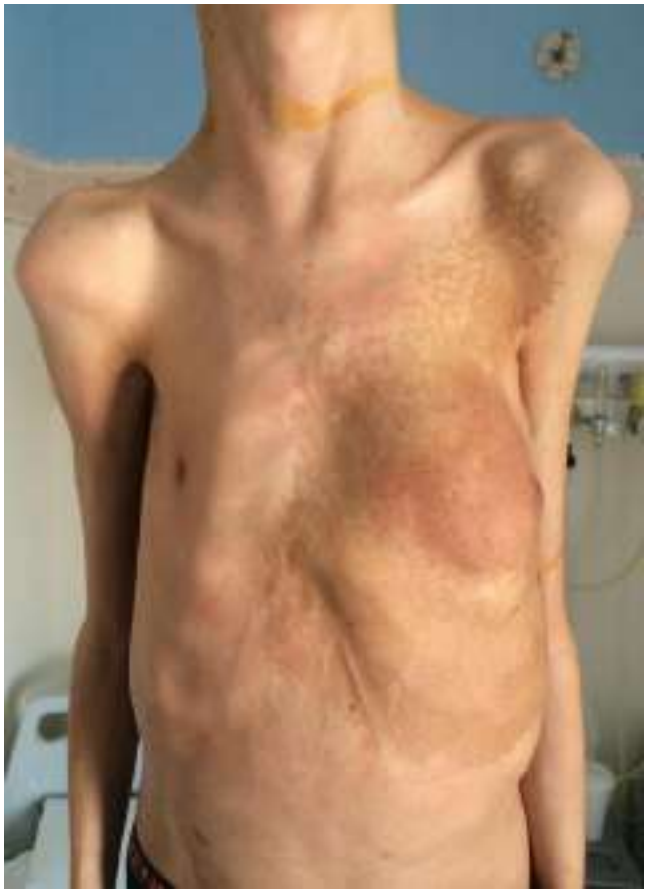

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Bone Lesions in Children with Neurofibromatosis

By Nikolaos Laliotis

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Chapter

Bone Lesions in Children with Neurofibromatosis

Nikolaos Laliotis

Abstract

Neurofibromatosis is often related with severe orthopaedic disorders in children. Bone lesions are rare but pose severe difficulties in management. It affects the spine and long bones. Lesions are associated either from enlargement of neurofibromas that affect the normal growth or from primary neurofibromatosis of long bones. Dystrophic scoliosis appears with short curves, with kyphosis and rotation of the apical vertebrae. Usually affect the thoracic spine, with penciling of the ribs. Surgical treatment is challenging in cases of rapid progression. Scoliosis may appear with curvatures similar to those in idiopathic scoliosis, without dysplastic changes of the vertebrae. Anterior bowing of the tibia is manifestation of NF and is distinguished from the benign posterolateral bowing. Evaluation of the medullary canal and presence of cystic lesions in the tibia is essential. Progression to pseudoarthrosis or pathologic fracture is common. Surgical management of tibial pseudoarthrosis remains a difficult procedure. Pseudoarthrosis may appear in fibula, radius or ulna but are extremely rare. Irregular eccentric bone cysts in long bones that are commonly diagnosed after a pathologic fracture, must be differentiated for NF. Malignant transformation of neurofibromas must be considered when there is rapid progression of the lesion.

Keywords: Scoliosis, dystrophic scoliosis, surgical management, spine in neurofibromatosis, spinal instrumentation, Congenital pseudoarthrosis tibia, fibula, radius, ulna, Idiopathic non-union tibia, fibula, radius, ulna, Neurofibromatosis tibia, fibula, radius, ulna

1. Introduction

Neurofibromatosis is a hereditary autosomal dominant disease associated with abnormal increase of neural cells, both from the central and peripheral nervous system. Children and adults are affected from the disease.

Orthopaedic manifestations of NF in children are found in the spine and the long bones. Alterations of the normal shape of the spine both in the frontal and sagittal plane appear, in the form of dystrophic and non-dystrophic scoliosis and kyphosis. It is unclear the exact mechanism for development of scoliosis in NF, as in general for scoliosis. Vertebral neurofibromas can erode the vertebrae either from the interior or the exterior, resembling congenital hemivertebra. Vascular and osteoblastic dysfunction may alter the shape of the vertebrae. Diagnosis of NF is based on the clinical criteria that include the dysplasia of long bones and spine lesions. Radiological evaluation both of x-rays and MRI and is important to properly follow the affected children. Management of dystrophic curves is a challenge for the pediatric spine surgeon.

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