



**Concept by Rigo**

## **Basic Level**

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**Unit 4: EOS, AIS, AdS**

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# Scoliosis: Classification according to the age of onset



Idiopathic Scoliosis is the most common form of morphological scoliosis and appears in apparently healthy children during any period of life

## IDIOPATHIC SCOLIOSIS

The prognosis, diagnosis, and operative indications related to curve patterns and the age at onset

J. I. P. JAMES, LONDON, ENGLAND

*From the Royal National Orthopaedic Hospital and the Institute of Orthopaedics*

THE JOURNAL OF BONE AND JOINT SURGERY

VOL. 36 B, NO. 1, FEBRUARY 1954

- Three peak periods of onset

- 0-3 years

- 5-8 years

- 10-end of growth



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- Three peak periods of onset

- 0-3 years.....INFANTILE

- 5-8 years.....JUVENILE

- 10-end of growth.....ADOLESCENT



## Revised Glossary of Terms

### SRS Terminology Committee and Working Group on Spinal Classification Revised Glossary of Terms

By the Working Group on 3-D Classification (Chair Larry Lenke, MD), and the Terminology Committee, March 2000

Chronological presentation of Idiopathic Scoliosis:

- 1) Infantile Scoliosis presenting from birth through age 2 + 11
- 2) Juvenile Scoliosis presenting from age 3 through age 9 + 11
- 3) Adolescent Scoliosis presenting from age 10 through the age of 17 + 11
- 4) Adult Scoliosis presenting from age 18 and beyond



## Early Onset Scoliosis (EOS)

- To confirm a worse prognosis in scoliosis beginning before the age of 10 in comparison with scoliosis beginning from 10 years of age and beyond, called Late Onset Scoliosis (LOS)

### PROGNOSIS IN IDIOPATHIC SCOLIOSIS

IGNACIO V. PONSETI and BARRY FRIEDMAN  
*J. Bone Joint Surg. Am.* 32:381-395, 1950.



## Early Onset Scoliosis (EOS) from Dickson R.A.

- EOS = Infantile Scoliosis and later on 'scoliosis present in children younger than 5 years of age, including Idiopathic, Neuromuscular, Congenital or Syndromic'

The Journal of Bone and Joint Surgery, British volume, Vol. 67-B, No. 2 | 1985 | 5

### Conservative treatment for idiopathic scoliosis

R.A. Dickson



EOS

LOS



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**EOS** is nowadays defined as curvature of the spine  $\geq 10^\circ$  in the frontal plane with onset before 10 years of age including congenital, neuromuscular, syndromic and idiopathic.



Scoliosis is a Biphasic Process and current diagnosis of EOS according to SRS means very little in terms of Natural History and Prognosis (Sebastik J 1996)

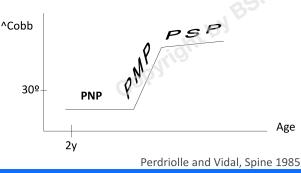
- EOS
  - It can go into progression before 10 years of age (Cobb angle at the end of growth used to be higher than  $90^\circ$ )
  - It can remain stable and progress only later during Adolescence With similar prognosis than AIS (or LOS)
  - It can spontaneously regress



## Natural History and Prognosis

- Age of ONSET is not so important but age of PROGRESSION

Period of Maximum Progression (PMP) occurs during rapid growth



Perdriolle and Vidal, Spine 1985



## Escoliosis sintomáticas. Causas.

Rebecca Sauvagnac Quera. MD  
PM&R specialist



### Structural Scoliosis are most idiopathic

- **Idiopathic (75%-80%)**
- Congenital (10%)
- Neurological (5%-7%)
- Recklinghausen (2%-3%)
- Others (Marfan, Beals, ... tumors)

Scoliosis Research Society



## Escoliosis congenital

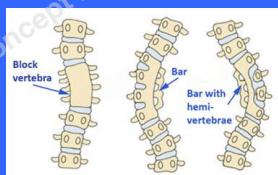
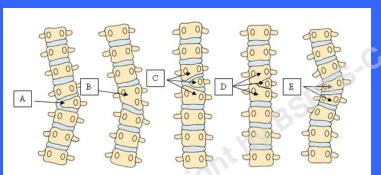
- Debido a un desarrollo vertebral anormal entre la 4a y 6a semana gestacional que resulta en un crecimiento asimétrico de la columna.
- Historia natural:
  - 25% no progresan
  - 25% progresan lentamente
  - 50% progresan rápido
  - Lo que determina la progresión curva: tipo de anormalidad, localización y edad del paciente
- Anomalías asociadas
  - Genito urinarias (20-40%)
  - Musculo esquelético
  - Cardiovascular (18-26%)
  - Anomalías del axis neural (40%)
  - Ex: VACTERL syndrome (vertebral anomalies, anorectale atresia, cardiac anomalies, tracheo-esophageal fistula and/or esophageal atresia, renal and limb anomalies); Klippel-Feil syndrome (cervical spine anomalies)

REVIEW ARTICLE. What's New in Congenital Scoliosis? Joshua M. Pahys, MD\* and James T. Guille, MD. J Pediatr Orthop 2016;00:000-000



## Escoliosis congenital

Deformidad causada por una formación anormal de las vértebras



Defect of formation

- A. Missplanned vertebra with scoliosis
- B. Misshapen vertebra fused to normal vertebra with scoliosis
- C. Three misshapen vertebrae without scoliosis
- D. Three fused vertebrae without scoliosis
- E. Trapezoidal shaped vertebrae with scoliosis

Defect of segmentation

<https://www.srs.org/professionals/online-education-and-resources/conditions-and-treatments/congenital-scoliosis>



## Escoliosis Neurológicas (1)

- Columna con una hipo o hiper tonía de los músculos del tronco con asimetrías de control de tronco
- En una columna en crecimiento
- Las deformidades de columna y tórax son indisociables → Los tratamientos son indisociables



## Escoliosis Neurológicas (2)

Table 1 Main neuromuscular etiologies of spinal deformity.

Central neurological causes	<b>Cerebral palsy</b>
Central motor neuron involvement	Hereditary ataxia (Friedreich, etc.) Syringomyelia Other central causes (encephalopathy, Rett's syndrome, etc.)
Peripheral neurological causes	<b>Acute anterio- postiomyelitis</b> Infantile spinal amyotrophy Hereditary motor and sensory neuropathy Hereditary sensory and vegetative neuropathy (familial dysautonomia)
Peripheral motor neuron involvement	
Mixed central and peripheral neurological causes	<b>Medullary lesion</b> Myelodysplasia Myelomeningocele
	Myasthenia
Neuromuscular junction (motor end-plate)	<b>Duchenne myopathy</b>
Muscular causes	Other muscular dystrophy Arthrogryposis

REVIEW ARTICLE. Neuromuscular scoliosis R. Vialle\*, C. Thévenin-Lemoine, P. Mary.  
Orthopaedics & Traumatology: Surgery & Research (2013) 995, S124–S139



## Escoliosis Neurológicas (3)

**Table 2** General population prevalence of scoliosis according to etiology [6].

Etiology	Prevalence (%)
Cerebral palsy	25
Myelodysplasia	60
Spinal amyotrophy	67
Friedreich's ataxia	80
Duchenne myopathy	90
Medullary lesion (< 10 years of age)	100

REVIEW ARTICLE. Neuromuscular scoliosis R. Vialle\*, C. Thévenin-Lemoine, P. Mary.  
Orthopaedics & Traumatology: Surgery & Research (2013) 99S, S124—S139



## Recklinghausen Neurofibromatosis type 1

- Enfermedad multisistémica
  - Cutáneos : tacas "Cafés au lait", Neurofibromas cutáneos
  - Extra cutáneos : Escoliosis, Pectus excavatum, otras deformidades esqueléticas... patologías cardio vascular
- 10% de los pacientes tendrán escoliosis (osteopenia y displasia) con
  - Scalloping of vertebrae
  - Bone dystrophy

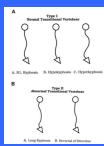


Neurofibromatosis type 1. Kevin P. Boyd et al. J Am Acad Dermatol. 2009 July ; 61(1): 1-16. doi:10.1016/j.jaad.2008.12.051.



## Marfan syndrome

- Enfermedades del tejido conectivo
- 62% de los paciente hacen escoliosis
- Patrón de curvas similar a los de las escoliosis idiopáticas, pero con diferencias de perfil



Sponseller PD, Hobbs W, Riley LH 3rd, et al. The thoracolumbar spine in Marfan syndrome. J Bone Joint Surg Am 1995;77(8):868; with permission.



## Others ...

- Otras patologías del tejido conectivo
  - Síndrome de Beals : Contracturas congénitas, aranodactilia, camptodactilia, kyphoscoliosis
  - Loeys-Dietz síndrome, Ehler Danlos síndrome...
- Tumoral : osteoma ostéoïde
- Infección : spondilodiscite



## Escoliosis sintomáticas

= Escoliosis secundarias =

Estas escoliosis tienen un doble potencial de progresión:

- El de la enfermedad. Cada tipo de escoliosis sintomáticas tendrá un evolución diferente.
- La misma que las escoliosis idiopáticas, durante el pico de crecimiento.



Escoliosis symptomaticas.  
Banderas rojas.

Rebecca Sauvagnac Quera. MD  
PM&R specialist



## Historia clínica

- Historia neonatal
- Desarrollo psicomotor
  - 3 meses Control de cabeza
  - 7 meses Se sienta solo
  - 9-17 meses Anda solo
  - Como los niños de su edad Del mismo curso escolar
- Historia médica personal:
  - Malformaciones asociadas genitourinarias, muscular esquelético, cardiovasculares...
  - Historia familiar de patologías neurológicas o óseas...



## Generalidades

- Insuficiencia respiratoria
- Soplido cardiaco
- Fiebre crónica
- Astenia, anorexia, slimming



## Valoración de la escoliosis

- Curva torácica izquierda
- « Rigid spine »
- Escoliosis angular



## Valoración Osteo articular

- Articulaciones: Rígidas o hyperlaxas
- Aranodactilia
- Camptodactilia
- Pectus



## Valoración neurológica

- Tono muscular : aumentado o disminuido
- Reflejos Osteo tendinosos (ROT) : ausentes, disminuidos o aumentados
- Reflejos cutáneo abdominales (RCA): Ausentes ou disminuidos
- Dismetría : pruebas dedo-nariz o talón-rodilla
- Marcha atáxica
- Movimientos anormales



# Valoración dermatológica

- Estigmas de anomalías del eje neural



- Tacas « café-au-lait »



- Neurofibromas



## Red flags

Congenital Scoliosis	Other malformation
Neurological scoliosis	<p>Neuro muscular (nerve, muscle...)</p> <ul style="list-style-type: none"> <li>- OTR Absence or diminution</li> <li>- Hipo tonia</li> <li>- Rigid spine</li> <li>- Joint stiffness</li> </ul> <p>Neuro central (cerebral palsy, syringomyelia...)</p> <ul style="list-style-type: none"> <li>- OTR Exacerbation</li> <li>- Hiper tonia</li> <li>- ACR absents</li> <li>- Joint stiffness</li> </ul>
Fridreich ataxia	<ul style="list-style-type: none"> <li>- Dismetria at finger-nose test, heel-knee test</li> <li>- Ataxic walking</li> </ul>
Medular lesion (syringomyela or occipito-cervical malformacion)	<ul style="list-style-type: none"> <li>- Left toracic curve</li> <li>- Asimetry of ACR</li> </ul>
Tumoral	<ul style="list-style-type: none"> <li>- Angular scoliosis</li> <li>- Alteración of general state</li> </ul> <p>- Escoliosis reactiva</p>
Marfan syndrome	<ul style="list-style-type: none"> <li>- Arachnodactyl</li> <li>- Joint hyperlaxity</li> <li>- Cutaneous hyperlaxity</li> <li>- Pectus excavatum</li> </ul>
Neurofibromatosis type 1 or Recklinghausen	<ul style="list-style-type: none"> <li>- Cafés au lait" macules, Cutaneous neurofibromas</li> <li>- Pectus excavatum</li> <li>- Heart murmur</li> </ul>

