Early Onset Scoliosis
EOS

Pierre LASCOMBES M.D.,
Nancy - France

Early Onset Scoliosis

1. Définition
2. Croissance et développement
3. Propositions thérapeutiques
4. Expériences et littérature
5. Discussion, propre expérience

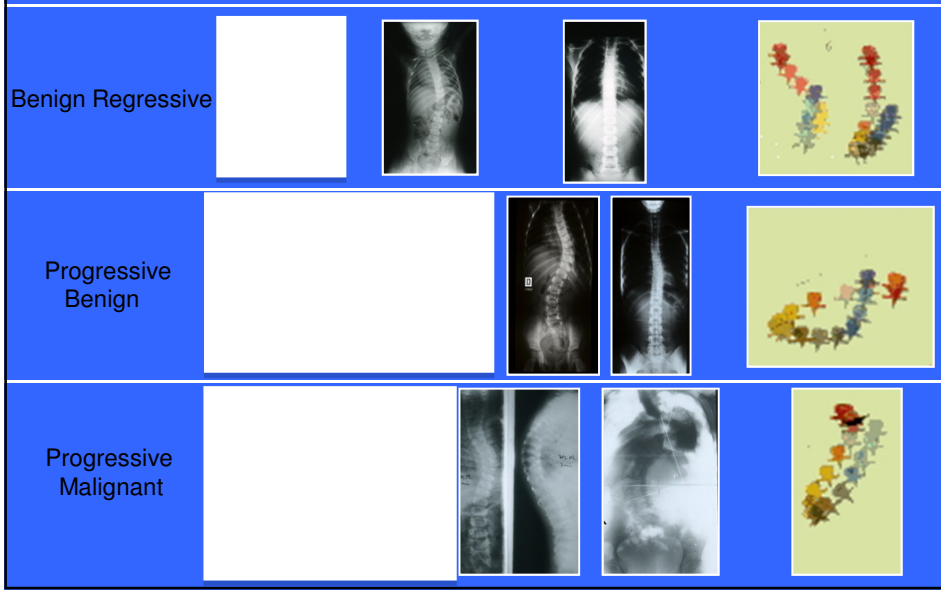
EOS : définition

- Scoliose idiopathique
  - Infantile : < 3 ans
  - Juvénile : 3 à 11 ans
  - Progression de la courbure ?
  - Risque de perturbation du développement pulmonaire chez l'enfant âgé de moins de 5 ans
- RA Dickson, 1994
  - Early Onset idiopathic Scoliosis = progressive curve under the age of 5 years

Infantile Idiopathic Scoliosis

0 – 3 Y of Age

Coll G Bollini

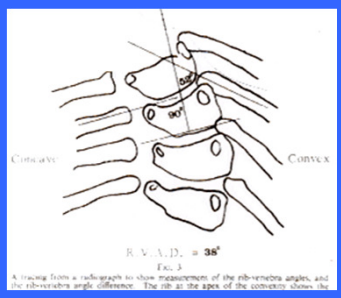


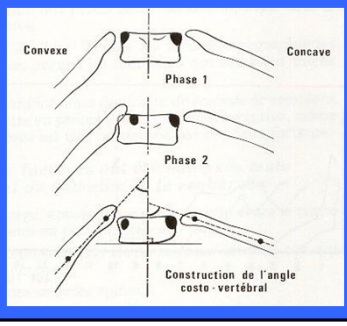
Courbe Progressive ?

Rib vertebral angle difference - RVAD (MH Mehta)

- < 20° = 80% régressive
- > 20° = 80% progressive

Phase 1 à Phase 2 (1974) : la tête costale côté convexe se superpose à la vertèbre apicale par rotation vertébrale





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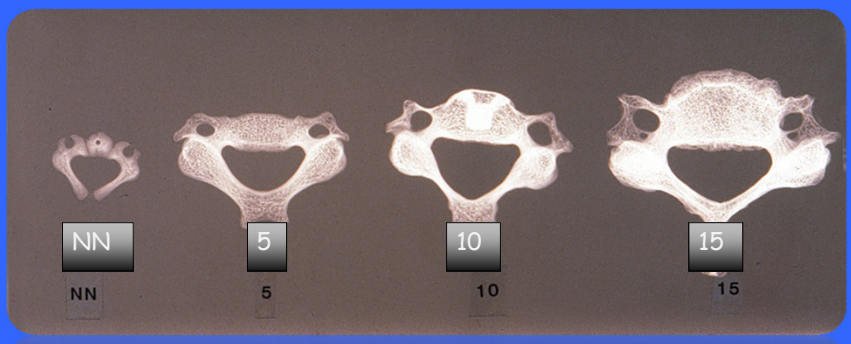
Trois challenges majeurs

1. Croissance du rachis
2. Croissance du thorax
3. Augmentation de la fonction pulmoniare

A Diméglio

Croissance coronale du rachis

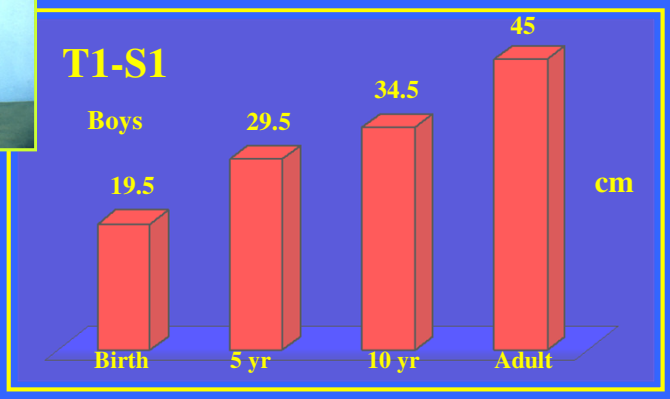


A l'âge de 5 ans, le canal rachidien atteint 95% de sa taille définitive

Une fusion circonférentielle ne peut pas entraînner un canal étroit

A Diméglio

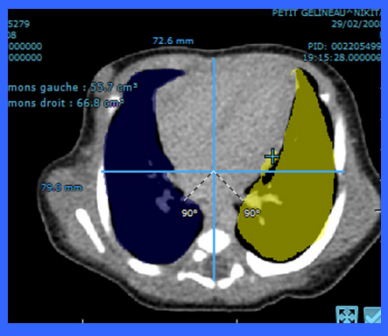
Croissance du rachis

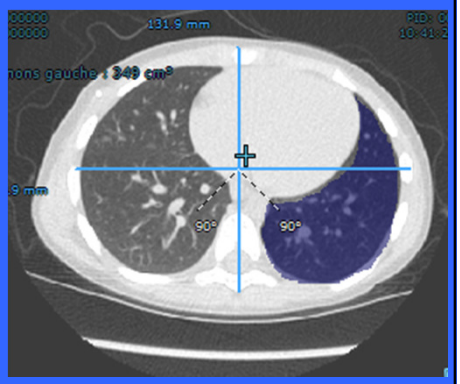


Une arthrodèses péri-vertébrale du segment T1-S1 à l'âge de 5 ans est responsable d'un déficit de la taille assise de 15 cm
(T1 – T12 =10 cm; L1 – L5= 5 cm)

A Diméglio

Croissance du thorax





Birth
AP: 72 mm
L: 79 mm
Volume Right 67 cm3
Volume Left 56 cm3

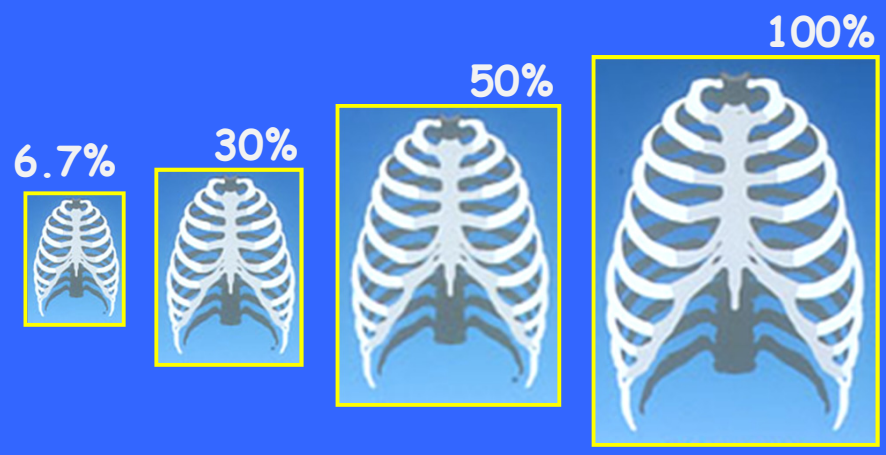
5 years
AP: 132 mm
L: 181 mm
Volume Right 398 cm3
Volume Left 349 cm3

Le diamètre frontal croit plus vite que le diamètre AP

A Diméglio

VOLUMETRIC GROWTH

The thorax: the fourth dimension of the spine

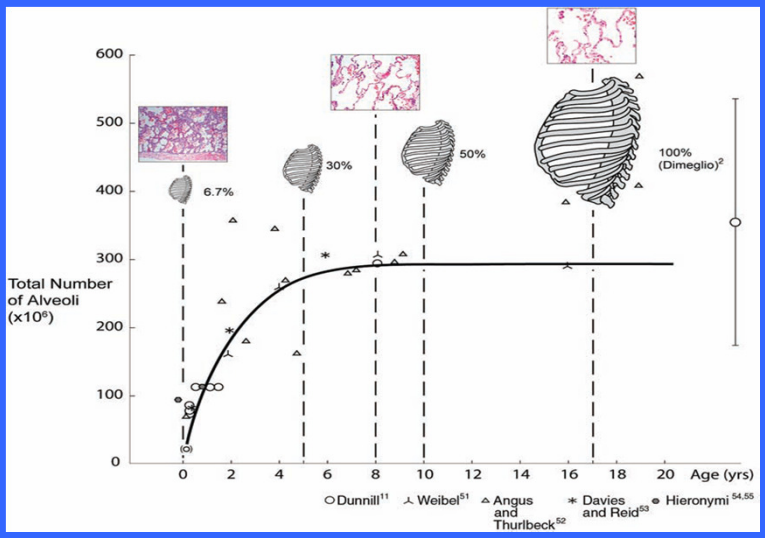


The growing spine, springer Velarg 1990

Arthrodèse du rachis et croissance du thorax

- F Canavese : une fusion postérieure T1-T6 (rat) diminue la croissance normale
  - Des vertèbres T1 à T6
  - Du sternum
  - De la cage thoracique dans son ensemble

Croissance alvéolaire pulmonaire



Déformation thoracique dans les scolioses sévères

- Thoracic insufficiency Syndrome

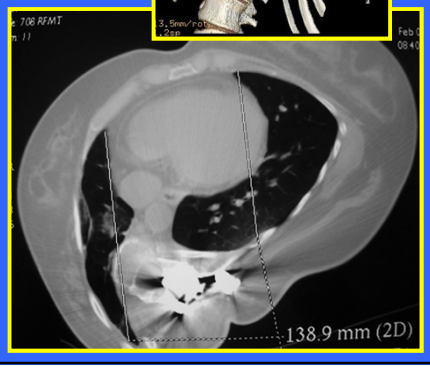
  Congenital scoliosis and fused ribs

  Campbell et al.
  J Bone Joint Surg (Am) 2003

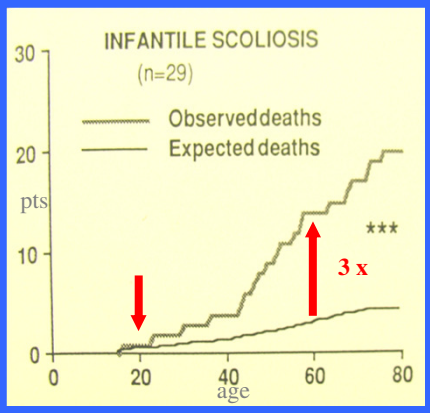
- Spinal penetration index

  Neuromuscular scoliosis
  Syndromes

  Dubousset et al.
  J Orthop Sci 2003



Capacité respiratoire et scolioses sévères



- Cobb > 110°
- CV < 43% normal

Pehrsson, Bake, Larsson, Nachemson, Thorax 1991

Pehrsson, Larsson, Oden, Nachemson, Spine 1992

Capacité respiratoire et scolioses sévères

- Carsets et plâtres, fusion après 10 ans
  - Capacité vitale (CV) = 68% de la normale
- Fusion vertébrale avant l'âge de 10 ans
  - Capacité vitale (CV) = 41% de la normale
    - (14% to 72%)

Respiratory Function and Cosmesis at Maturity in Infantile-onset Scoliosis Goldberg, C. J.; Gillic, I.; Connaughton, O.; Moore, DP Fogarty, Canny, G. J. Dowling, F. E. Spine Volume 28(20), 2003.

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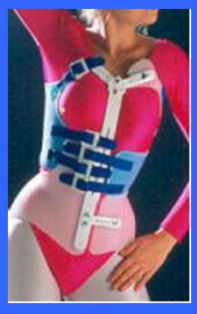
Traitement initial : plâtres et corsets



Résultats des plâtres et corsets

MH Mehta (JBJS Br; 2005)

- 35% des enfants > 30 mois nécessitent une fusion vertébrale à l'âge de 10 ans 4 mois
- Intérêt du halo crânien avant un plâtre
- Traitement d'attente de la fusion

Instrumentation rachidienne sans greffe 1978 - John Moe

Harrington instrumentation without fusion combined with the Milwaukee brace for difficult scoliosis problems in young children. Orthop Trans.1979;3:59

J. Pediatr. Orthop., 1997, 17, 734-742

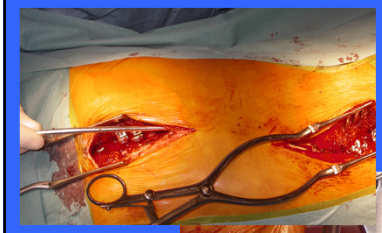
- 67 children : 31 neuromuscular
- Cobb angle : initial = 67° , final = 47°
- Growth = 3.1 cm (FU = 3.1 years)
- Growth = 0.08 cm / segment / year
- Complications :
  - Hook dislocations : 21
  - Rod fractures : 12

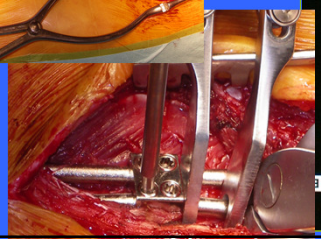
Spinal Instrumentation Without Fusion for Progressive Scoliosis in Young Children

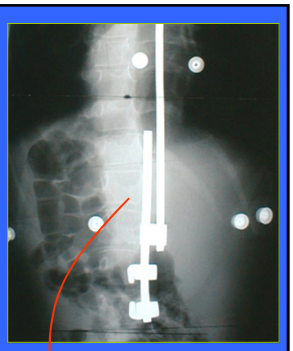
LTC William R. Klemme, MC, USA, *Francis Denis, M.D., *Robert B. Winter, M.D., *John W. Lonstein, M.D., and †Steven E. Koop, M.D.

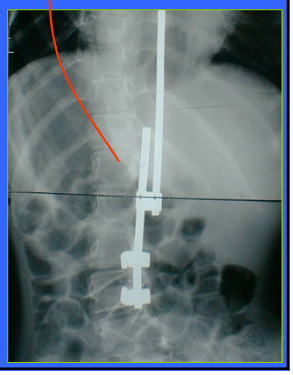
Expérience de la « SQR »

- pinces vertébrales
- greffe des appuis
- cintrage sagittal de la tige

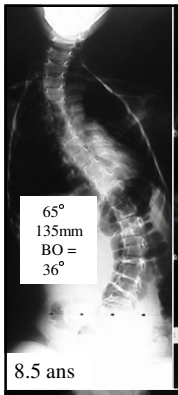


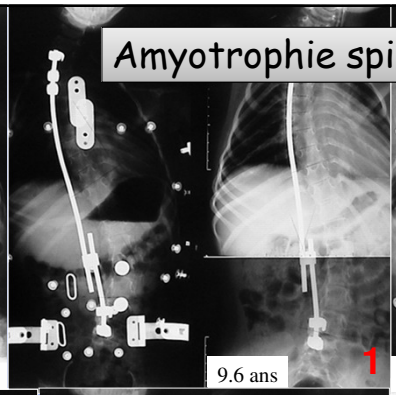


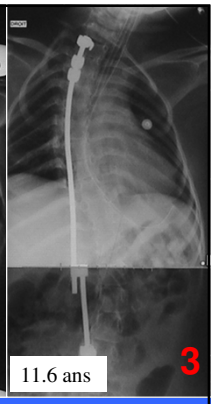


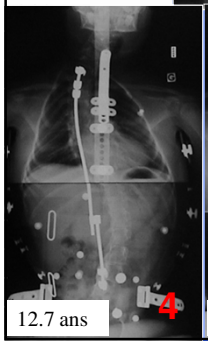


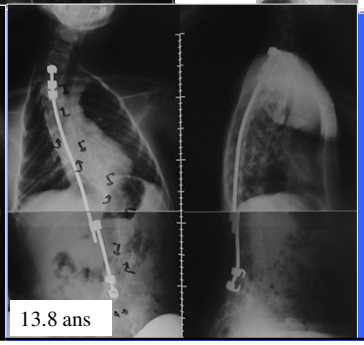
Amyotrophie spinale





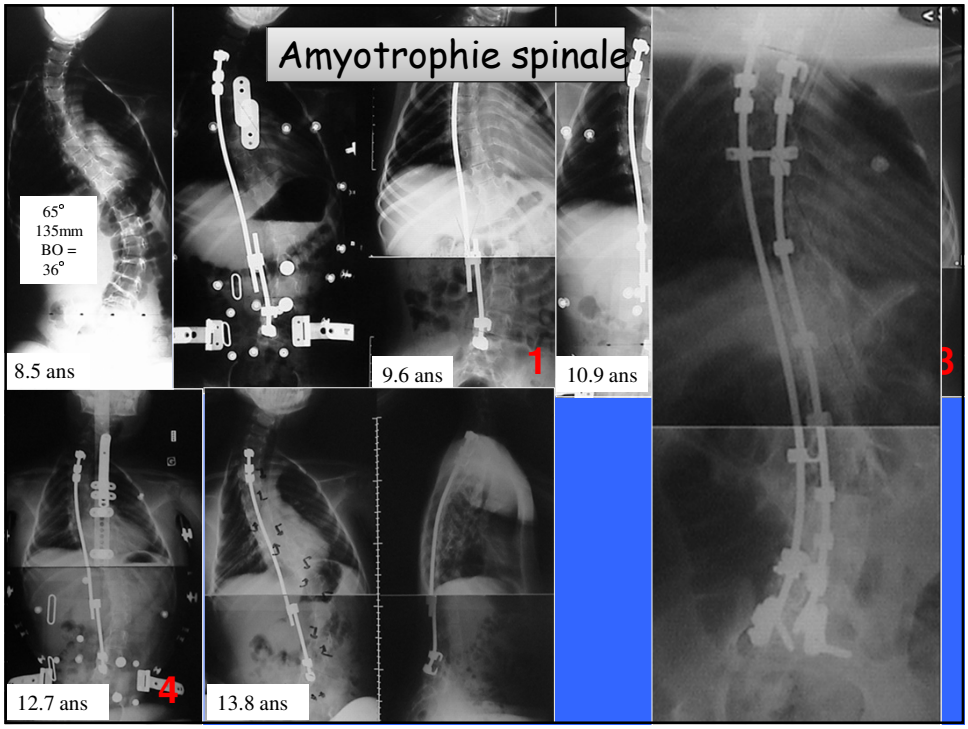






Au bout de 5 ans :

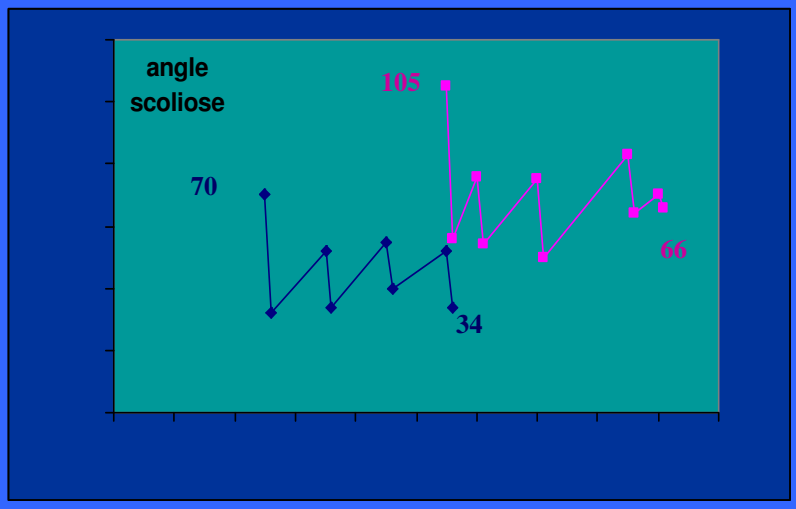
2 tiges sous faciales
1 domino
4 allongements
1 arthrodèse



Expérience de Nancy - SoFCOT 2005

- Amyotrophie spinale infantile : 12 cas
- Angle de Cobb initial moyen: 60,3° (41 - 86°)
- Nombre moyen d'allongement: 3,5 (2 - 6)
- Délai moyen : 9 mois (6 - 23)
- Gain angulaire moyen par allongement : 12,4° (0 -15,8°)
- Gain moyen en longueur par allongement : 10,6 mm
- Gain total en longueur d'allongement : 39,1mm (0-89)
- Angle de Cobb moyen au recul maximal : 40,8° (29 - 65°)
- Complications post-opératoires: 3 cas
  - un sepsis après un 6ème allongement
  - une complication mécanique (crochet proximal)
  - un glissement de domino (échec d'allongement)

2 exemples

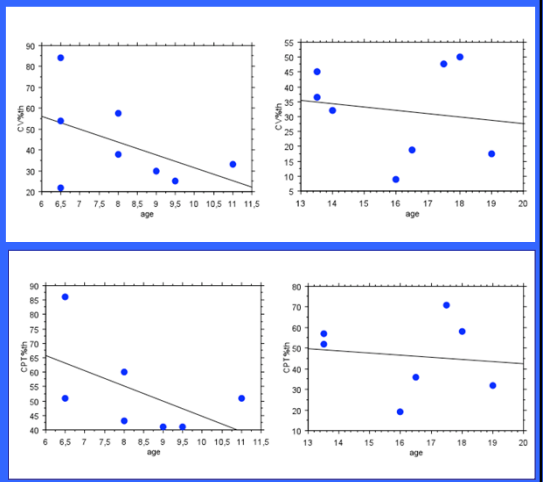


Effets de la « SQR » sur la fonction pulmonaire (SoFCOT 2006)

Pre - op Post - op

Pulmonary vital capacity

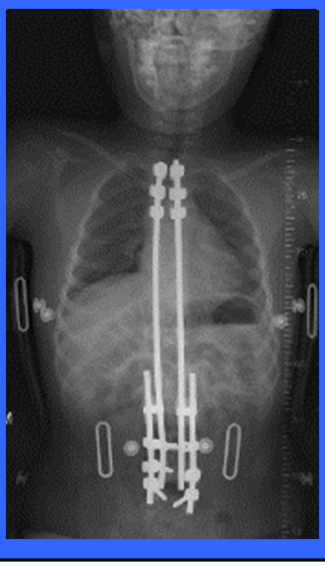
Total pulmonary capacity

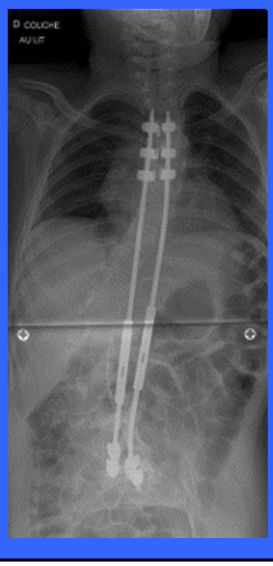


SQR stoppe la diminution de la fonction respiratoire dans l'amyotrophie spinale

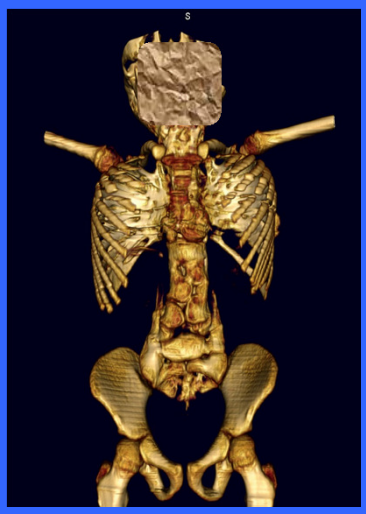
P Lascombes

De la tige sous fasciale, avec greffe des extrémités aux « tiges de croissance »





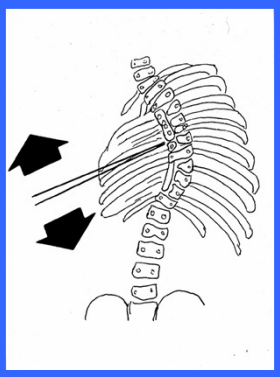
Natural history model of early spine fusion: Jarcho-Levin Syndrome

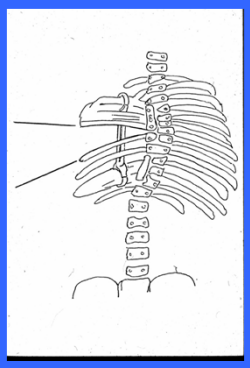


- 20 patients
- Taille assise = 24,2% / N
- CV = 27,9% / normal

Ramirez, et al. JBJS, 2007

VEPTR Opening Wedge Thoracostomy for Fused Ribs /Scoliosis





-27 pts

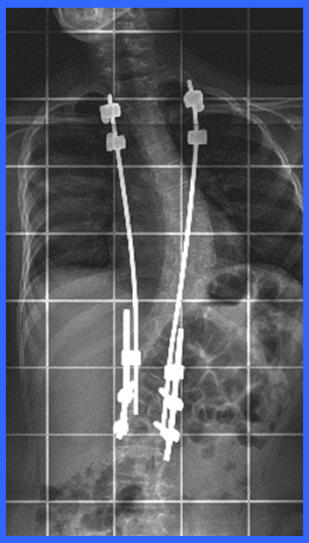
-Mean 25º correction

- JBJS 2004

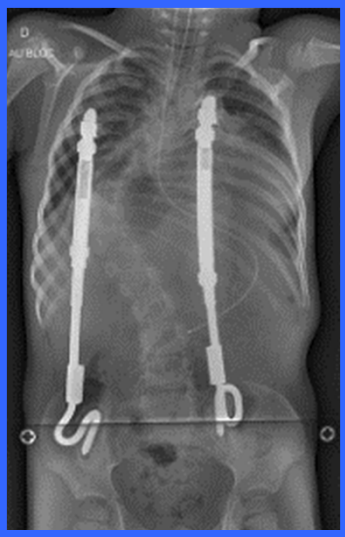
R Campbell

Principe de l'expansion de la cage thoracique

Rib distractor - Medtronic



VEPTR - Synthes



Shilla growing rod – RE McCarthy

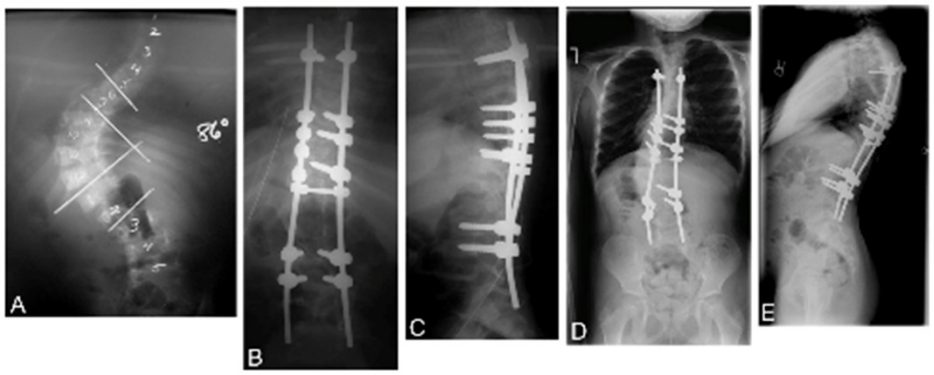
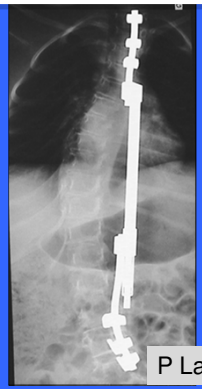


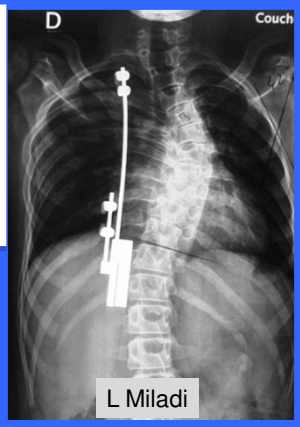
Figure 3. **A,** Preoperative radiograph of a 3 year old girl with a severe left thoracic idiopathic curve (Courtesy of R. E. McCarthy, MD[40]). **B, C,** Immediate postoperative correction using guided growth. The apical area has been corrected and fused by anterior release and posterior pedicle instrumentation T8–T11. The rods are left long and are not fixed to the T5 anchors proximally and the L2 and L3 anchors distally. **D, E,** 4½ years later, there has been growth at both ends of the construct, as the nonlocked screws have migrated along the rods. The apical deformity remains corrected.

In : CE Johnston, Spine 2010

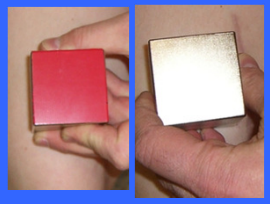
Fusionless programme

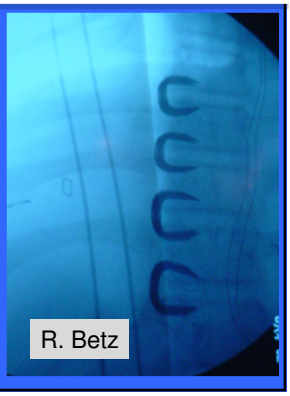


P Lascombes



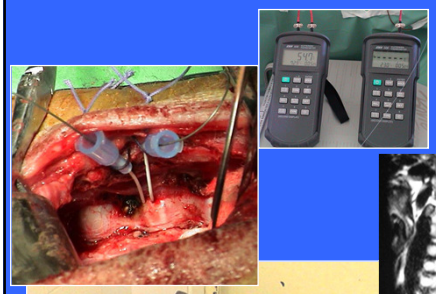
L Miladi

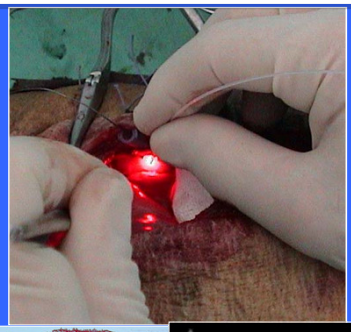


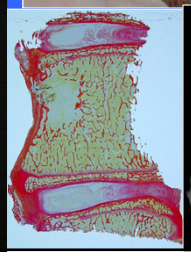
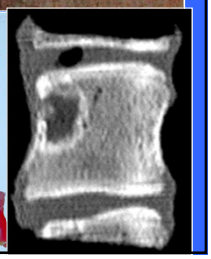


R. Betz

Fusionless programme
G Bollini – P Lascombes





CL Scoliosis, 7 y.o.
Congenital diaphragmtic hernia
Lung dysplasia

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BA Akbarnia M.D.

Table 1. Comparison of Indications, Treatment, and Complications in GR and VEPTR in EOS

	Growing Rods	VEPTR
Best indication	Normally segmented spine, flexible chest deformity	Thoracogenic scoliosis or fused ribs
Relative contraindication?	Primary chest wall deformity	Poor soft tissue coverage
Multiple operations needed?	Yes	Yes
Upper thoracic kyphosis?	Possible control	Poor control
Spine growth?	+	+
Chest deformity correction?	When flexible	Direct, invasive
Ease of final fusion	Difficult, scarred	Easier, unscarred
Final fusion needed?	Yes	Yes
Failures—common	Rods break	Rib attachments drift
Complication—severe	Spontaneous posterior spine fusion	Chest wall stiffness

GR indicates growing rods; VEPTR, vertical expandable prosthetic titanium rib; EOS, early onset scoliosis.

Meilleure indication = rachis normallement segmenté (EOS)

BA Akbarnia M.D.

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Cyphose thoracique proximale = mauvais contrôle avec VEPTR

BA Akbarnia M.D.

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Failures—common	Rods break	Rib attachments drift
Complication—severe	Spontaneous posterior spine fusion	Chest wall stiffness

GR indicates growing rods; VEPTR, vertical expandable prosthetic titanium rib; EOS, early onset scoliosis.

Facilité de la fusion = Difficile avec GR (cicatrice, fibrose)

BA Akbarnia M.D.

Table 1. Comparison of Indications, Treatment, and Complications in GR and VEPTR in EOS

	Growing Rods	VEPTR
Best indication	Normally segmented spine, flexible chest deformity	Thoracogenic scoliosis or fused ribs
Relative contraindication?	Primary chest wall deformity	Poor soft tissue coverage
Multiple operations needed?	Yes	Yes
Upper thoracic kyphosis?	Possible control	Poor control
Spine growth?	+	+
Chest deformity correction?	When flexible	Direct, invasive
Ease of final fusion	Difficult, scarred	Easier, unscarred
Final fusion needed?	Yes	Yes
Failures—common	Rods break	Rib attachments drift
Complication—severe	Spontaneous posterior spine fusion	Chest wall stiffness

GR indicates growing rods; VEPTR, vertical expandable prosthetic titanium rib; EOS, early onset scoliosis.

Complications : GR = ruptures de tiges
VEPTR = arrachage costal

BA Akbarnia M.D.

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Table 1. Comparison of Indications, Treatment, and Complications in GR and VEPTR in EOS

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GR indicates growing rods; VEPTR, vertical expandable prosthetic titanium rib; EOS, early onset scoliosis.

GR = postérieure fusion ± spontanée
VEPTR = rigidité de la cage thoracique

JS Yang, JPO; 2010 – Growing Spine Study
Group : Surgical procedures indication

- Progressive curves 73 ± 20°
- Mean age 6 ± 2.5 years
- Dual rods > single rod
- Lengthening interval = 8.6 months

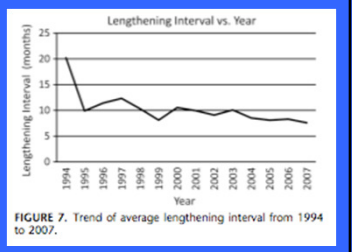
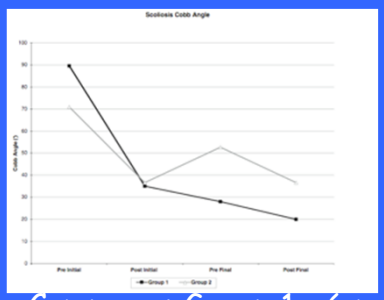
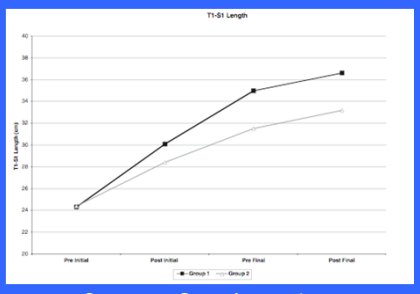


FIGURE 7. Trend of average lengthening interval from 1994 to 2007.

Effect of frequency of the legthening
BA Akbarnia, Spine; 2008





Compare : Group 1 ≤ 6 mois versus Group 2 > 6 mois
A corréler avec les risques infectieux

Law of diminishing return (WN Sankar, Spine) T1-S1 gain
- 1st lengthening = 1.04 ± 0.72 cm
- 7th lengthening = 0.41 ± 0.58 cm (p=0.007)

Chest wall expansion : VEPTR

Résultats & Complications (CC Hasler, Eur Spine J; 2010) – 23 enfants

- 23 complications
  - 10 problèmes cutanés
  - 5 dislocations d'implants
  - 2 ruptures de tiges
  - 6 infections
- Résultats
  - Cobb : 68° (10 – 110°) à 51° (0 – 105°) @ FU
  - Bassin oblique : 33° (13 - 60°) à 16° (0 – 42°)

Risque neurologique

WN Sankar, Spine; 2009 : 782 GR chirurgies

- Modification du monitoring
  - 0.9% implantation / modification des implants
  - 0.5% allongement
- Action prise: diminution de la correction, changement d'une vis

DL Skaggs, JBJS Am; 2009

- Plexus brachial = post VEPTR

Bilan pré-opératoire
CE Johnston, Spine; 2010

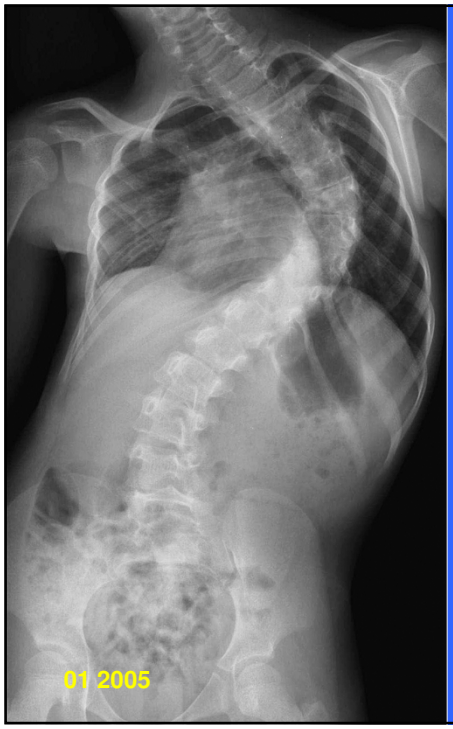
- Evaluation et préparation pulmonaire
- IRM médullaire (idiopathique ?)
- Bilan nutritionnel et prise en charge
- Evaluation de l'ostéopénie

Early Onset Scoliosis

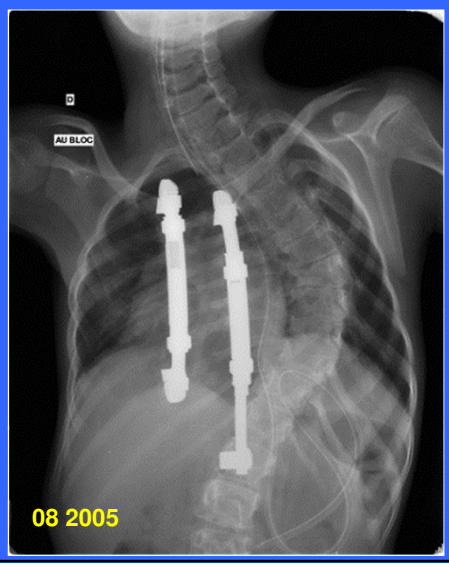
1. Définition
2. Croissance et développement
3. Propositions thérapeutiques
4. Expériences et littérature
5. Discussion, propre expérience

Cas # 1

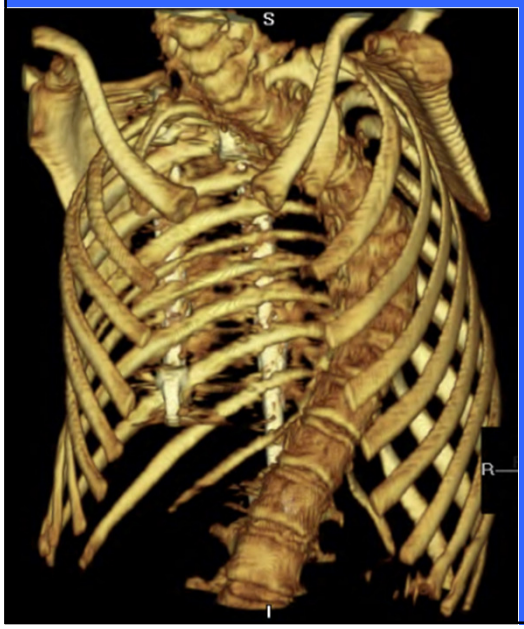
CL Scoliosis, 7 y.o.
Congenital diaphragmtic hernia
Lung dysplasia

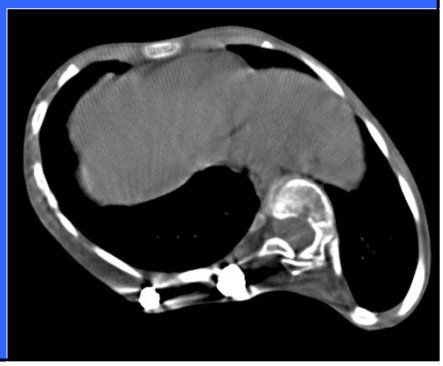


- VEPTR
  - between ribs # 3 & 10
  - Between rib # 4 and L1

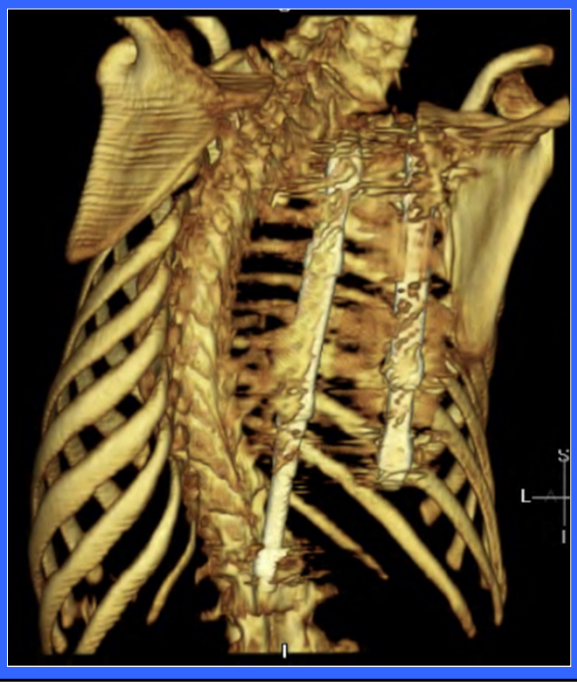


8.5 y.o.



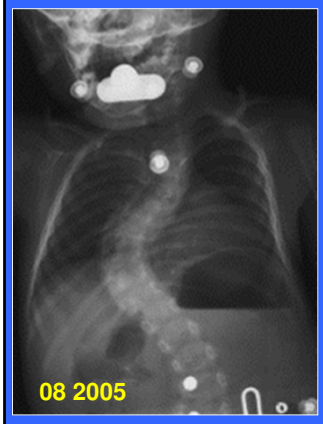


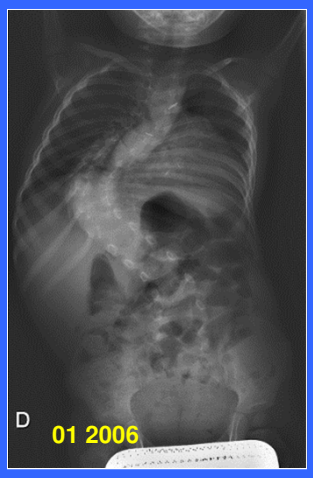
- Passed away age 10, due to respiratory failure…



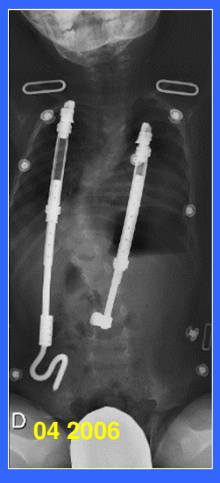
Cas # 2

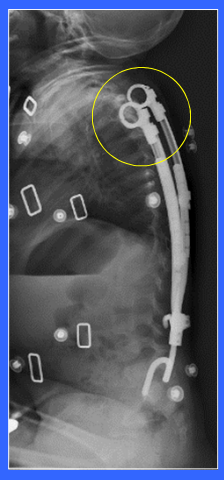
Garçon, 3 ans, « scoliose infantile », 22q11

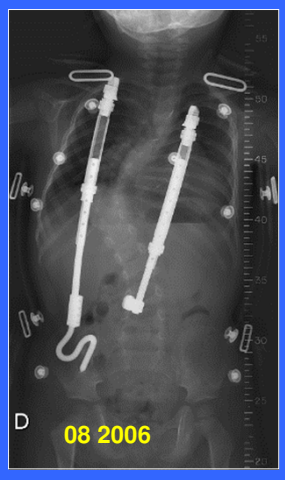


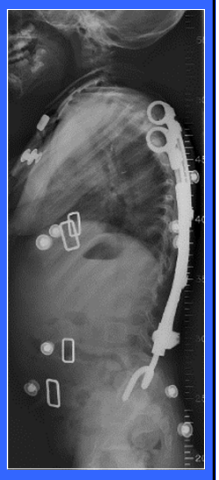


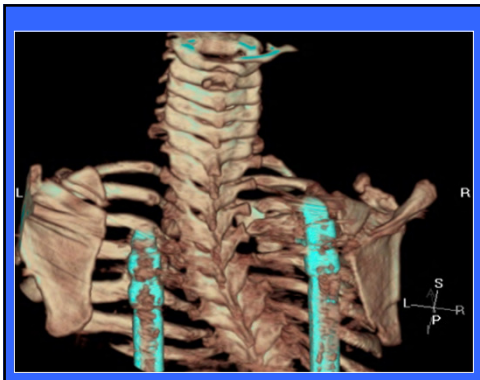
But : 2 VEPTR costo – iliaque bilatéral



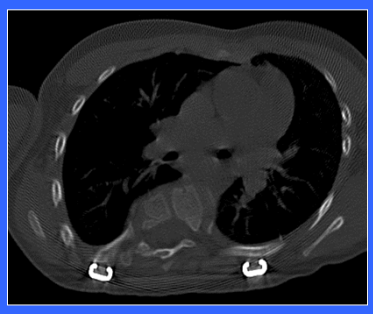


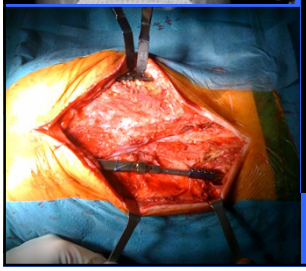




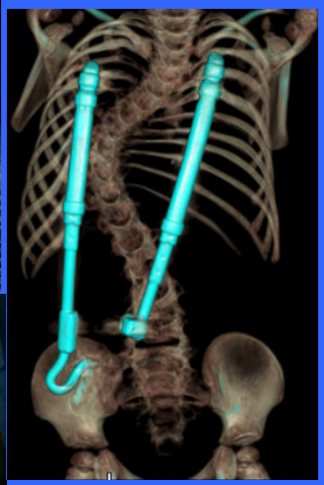


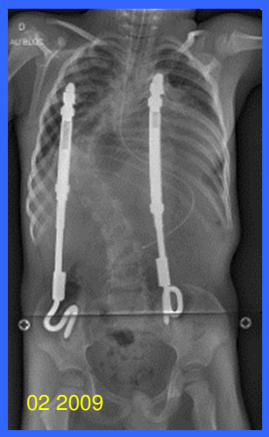
Août 2006:
- perte d'appui 4ème côte
- bon équilibre avec les deux tiges

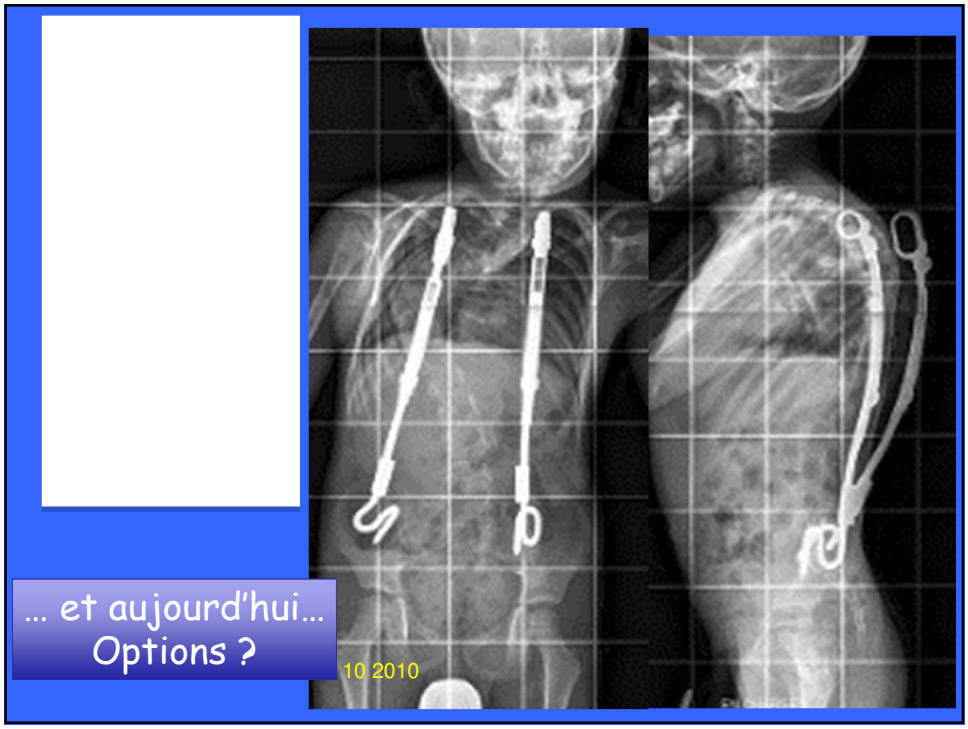




Changement du distracteur droit

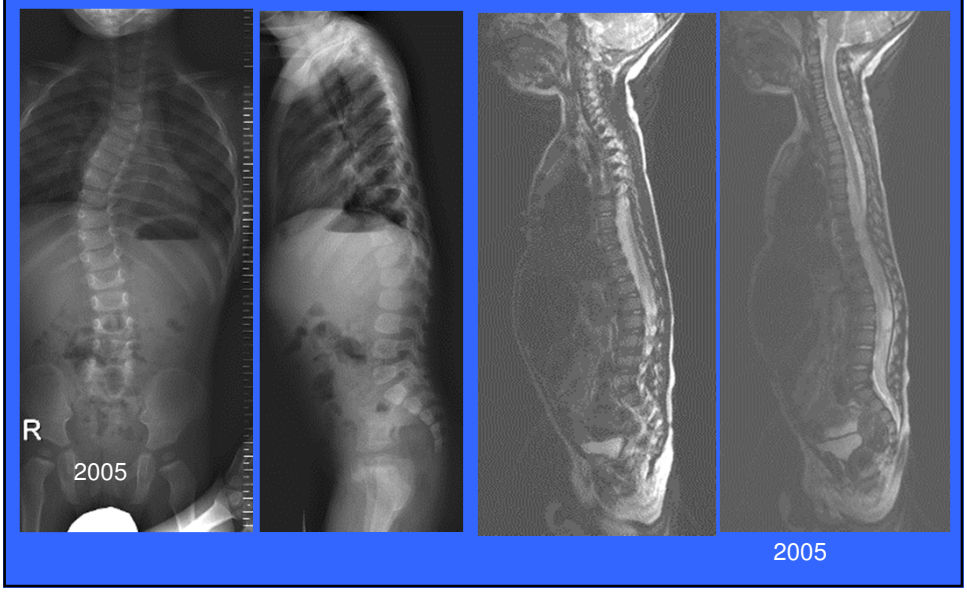






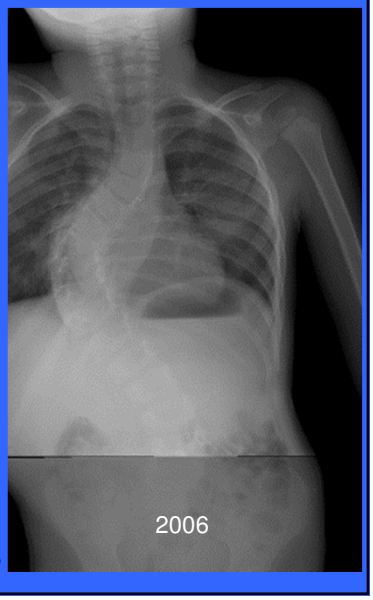
Cas # 3

Thomas L, garçon, né en 2003 : Neurofibromatose



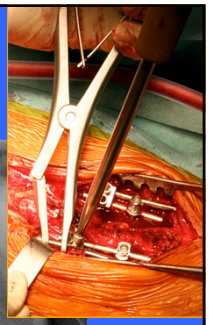
Boy, born 2003 : Neurofibromatosis

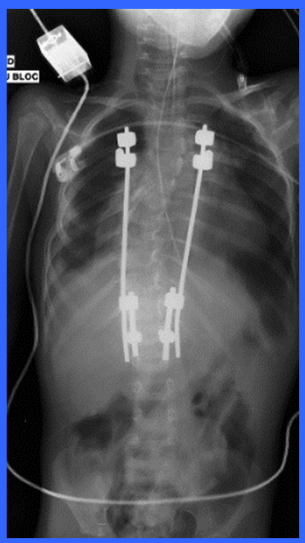
June 2005 = 30°
October 2006 = 75°

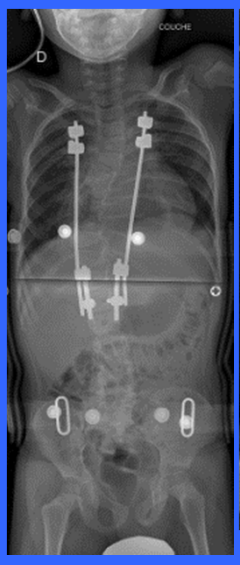


Halo gravity and anterior thoracic release

L1 – Rib distraction / 1st Lengthening

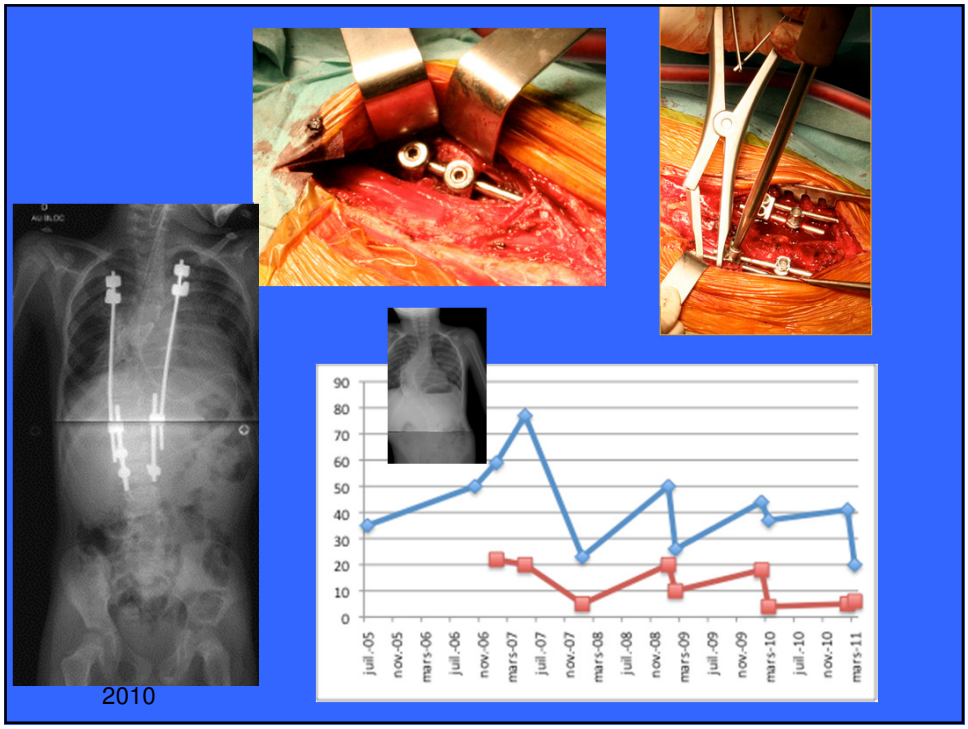






2008

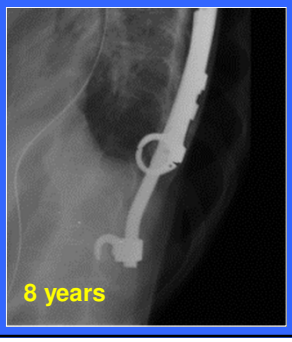
2009

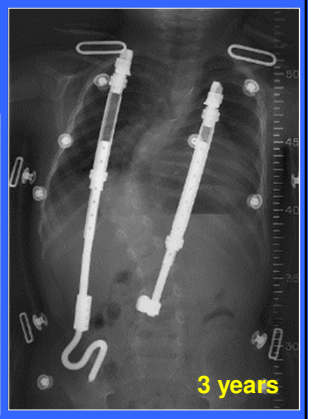


2010

1. Taille des implants

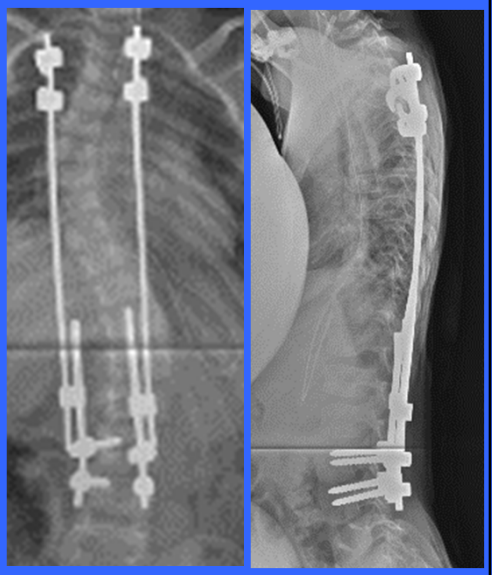
- Avant 3 ans, le « VEPTR » est gros
  - Fixation costale
  - Crochets laminaires
  - Fixation iliaque





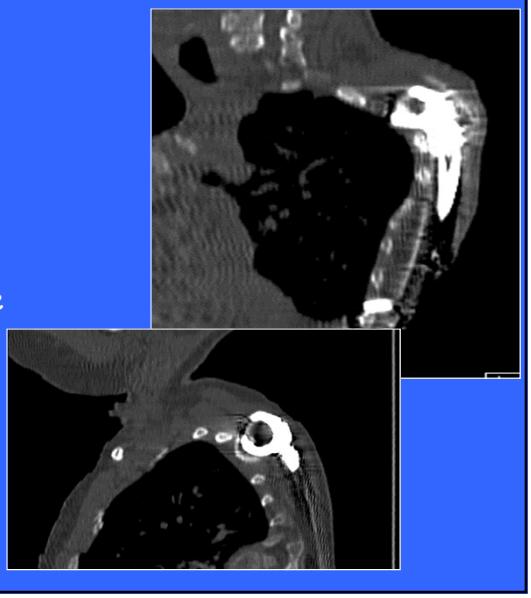
2. Appuis distal rachis

- Double vis pédiculaires
- Fixation pelvienne = ILIAQUE



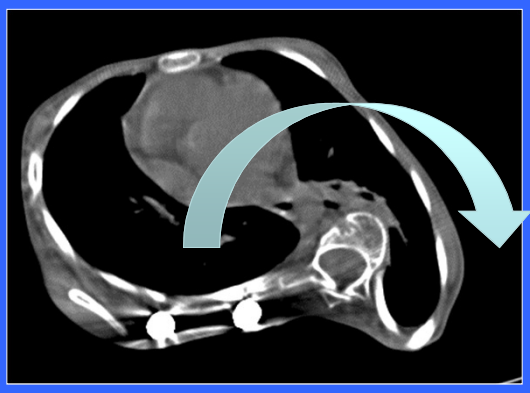
3. Arrachage d'appui costal et cyphose

- Même si l'appui est extrapériosté
- Induit par la rigidité de l'appui lombaire ou iliaque
- Cyphose sus-jacente

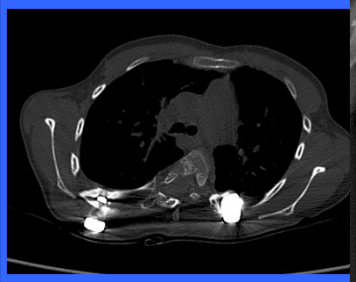


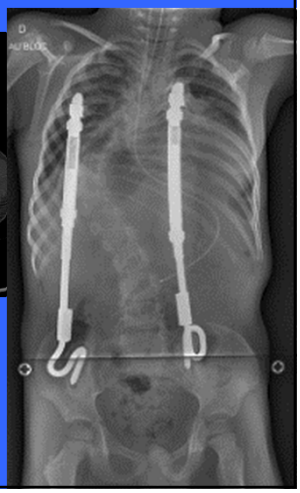
4. Rotation vertébrale

- La distraction concave augmente la rotation
- Les implants poussent vers l'avant



L'appui convexe équilibre le plan horizontal

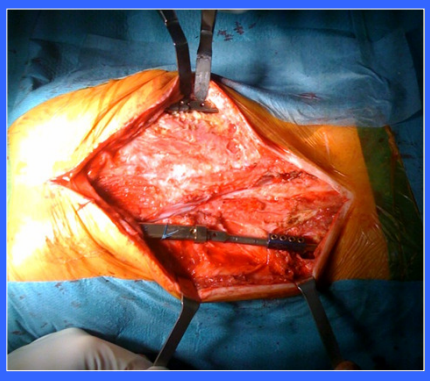




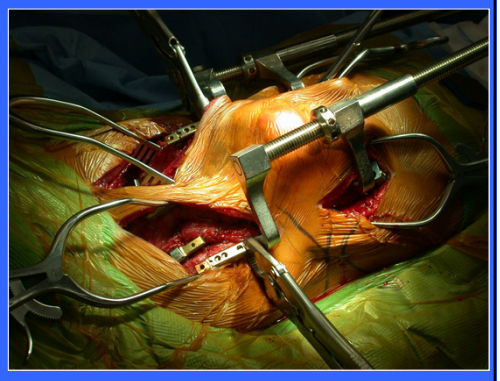
5. Stiff or not stiff ?

- Tige proximale : 3.5 mm
- Tige distale : 4.5 mm
- Arrachage costal proxi = 0
  (Nancy series – Eva Polirstock)
- « Flexible rib distractor » ?

6. Incision médiane postérieure

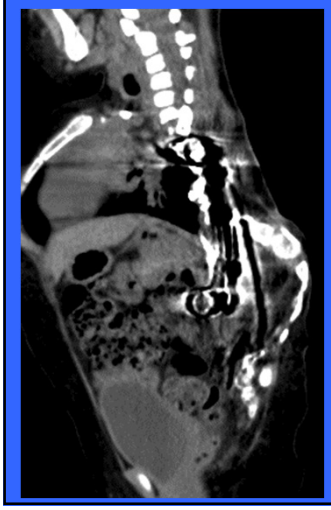


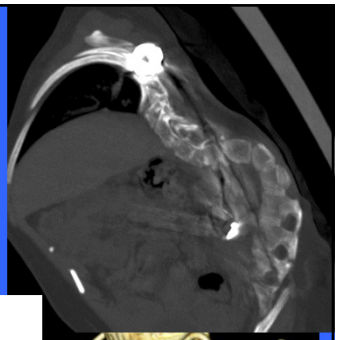
P Lascombes

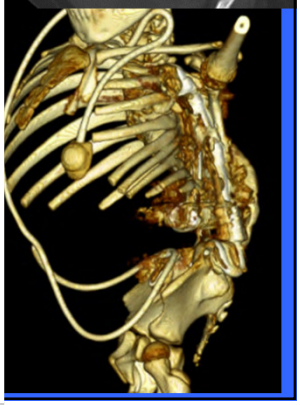


R Campbell

Fille 8 ans, myelomenigocèle niveau T12, opérée 17 fois ailleurs….

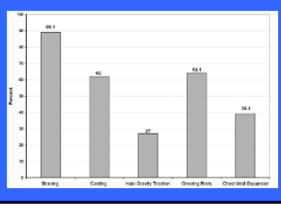






Traitement actuel POSNA

- 1er Scénario : 2 ans, 50° progressive, RVAD > 20° , phase 2
  - 55.7% = plâtres - corsets
  - 32.9% = corsets
- 2ème Scénario : 5 ans, 70° progressive
  - 52.9% = growing rod construct
  - 10.1% = chest wall expansion device
- Halo = 27%



CL, né en 1998

2001 = 20° , 2003 = 30° , plâtres & corsets

2007

Deux tiges de croissance,
Legacy 4.5 mm

Conclusion
Distraction costale

1. Gagner du temps sur la rigidité du rachis / GR
2. Débuter à 50° Cobb – Halo si > 80°
3. Double tiges – droite et gauche
4. Solide fixation inférieure (vis – iliaque)
5. Fixation costale « élastique »
6. Si cyphose = halo, libération antérieure et fixation haute T1 – T2
7. Abord sous fascial toujours médian

MERCI