

# SCOLIOSES CONGENITALES

Gerard BOLLINI Hôpital d'Enfants de la Timone Marseille



Recent evidence from work performed in fish, chick and mouse embryos indicates that **segmentation of the embryonic body relies on a molecular oscillator called the segmentation clock**, which requires Notch signaling for its proper functioning.

**In humans, mutations in genes required for oscillation, such as Delta-like 3 (DLL3), result in abnormal segmentation of the vertebral column**, as found in spondylocostal dysostosis syndrome, suggesting that the segmentation clock also acts during human embryonic development.



**Pourquie, O 1; Kusumi, K 2**

**Clinical Genetics. 60(6):409-416,  
December 2001.**





## Hemivertebra in Monozygotic Twins

Four cases have been reported in which one identical twin had a congenital scoliosis and the other did not. In two of these cases the second twin had an idiopathic scoliosis, and in two the spine of the second twin was normal.

Akbarnia et al reported on identical twins, one with congenital scoliosis and the other with congenital kyphoscoliosis. Both had multiple congenital vertebral and rib abnormalities.

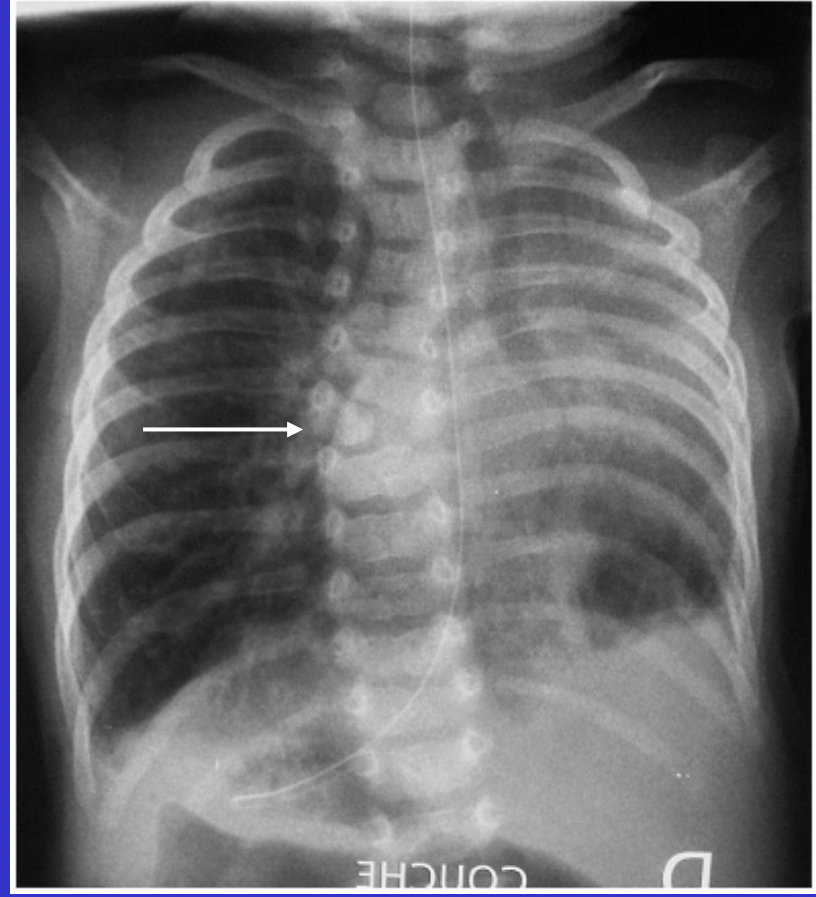
Only one case of **similar vertebral anomalies in twins exists in the literature**. Haffner reported on a pair of congenital twins with lumbar hemivertebra. We are presenting a case of monozygotic twins, each with a single thoracic hemivertebra.

Sturm, Peter F. MD,\*; Chung, Raymond  
MD,† and; Bomze, Shira R. RNFA\*

SPINE Volume  
26(12) 15 June  
2001 pp 1389-1391

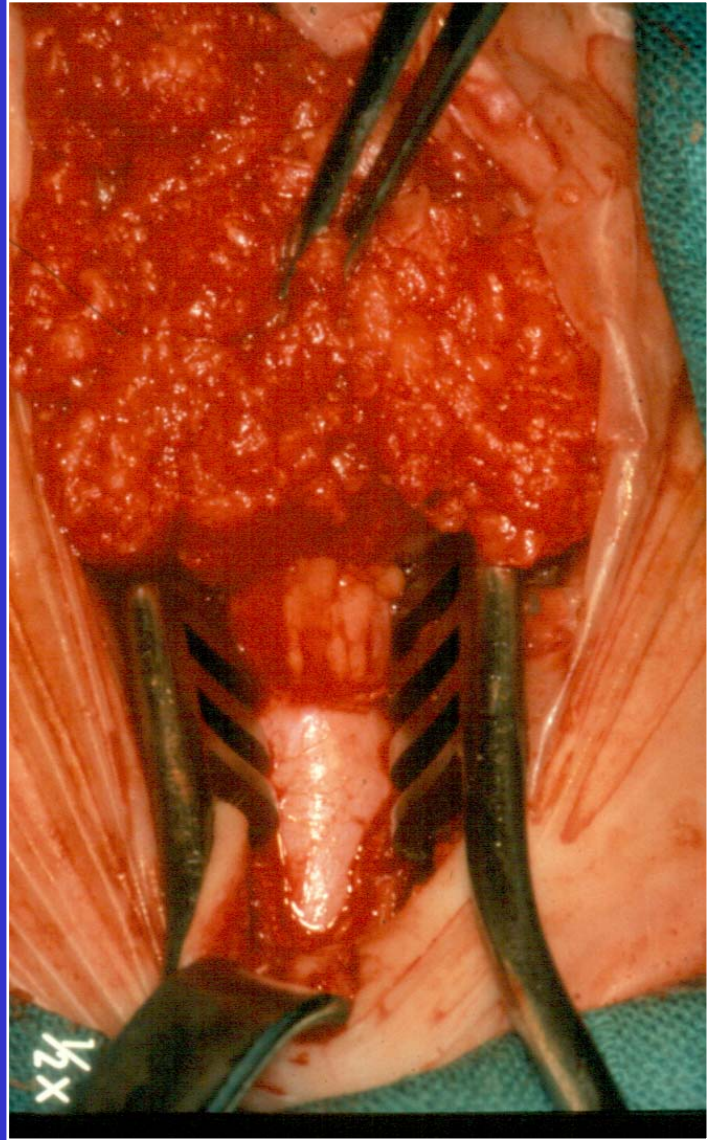
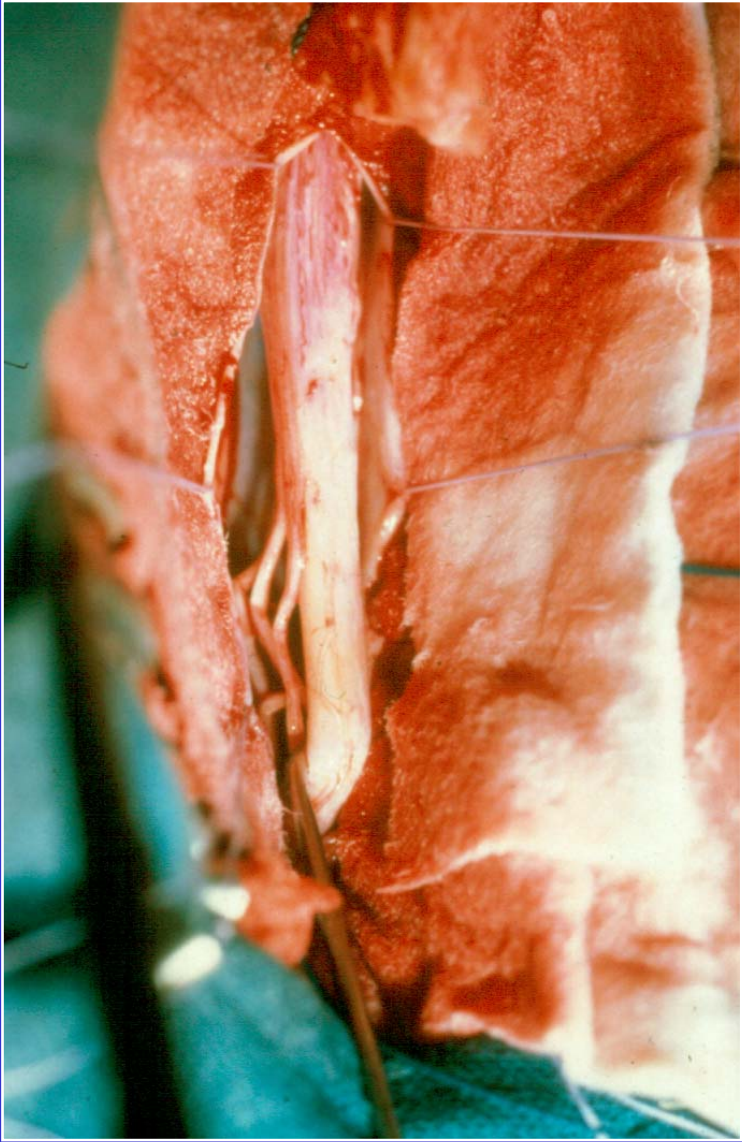












## Scolioses congénitales et Dysraphisme Spinal fermé

	<b>Nombre de patients</b>	<b>Anomalies Intracanales</b>
<b>Elsebaie 2003</b>	126	46 ( <b>37%</b> )
<b>Mac Master 1984</b>	251	46 ( <b>18.3%</b> )
<b>Suh 2001</b>	41	13 ( <b>34%</b> )
<b>Prahinski 2000</b>	30	9 ( <b>30%</b> )

**La découverte d'une malformation congénitale du rachis à la naissance implique;**

- la réalisation d'une échographie du système urinaire**
- La réalisation soit d'une échographie rachidienne en période néonatale soit d'une IRM vertèbro-médullaire ensuite**



Five patterns of deformity were recognized.

Hemivertebra was the most common type, and unilateral unsegmented bar with contralateral hemivertebra was the most severe and most progressive pattern of deformity.

**The curves measured  $>40^\circ$  in 70% of the patients who had reached maturity.**

**The curve progression index was  $9^\circ$  for unilateral unsegmented bar with contralateral hemivertebra, and  $6^\circ$  without contralateral hemivertebra.**

**This index was  $1.5^\circ$  for hemivertebra and complex type of deformity, and  $0.5^\circ$  for block vertebra.**

Unbalanced fully segmented hemivertebra was next after the two types of unsegmented bars in terms of potential for progression. Presence of fused ribs on concave side of lower thoracic curves increased the rate of curve progression.

### **Patterns and Progression in Congenital Scoliosis**

Shahcheraghi, G. Hossain M.D., F.R.C.S.(C); Hobbi, M. H. M.D.\*

SPINE Volume 19(6)

November/December 1999

p 766

**LORDOSE**

Barre postérieure

Barre postero-latérale

Barre latérale

**SCOLIOSE**

Bloc complet

Vertèbre binucléée T-L

Ossification ant. Prog.

Barre antérieure

Agénésie corp. Ant.

HVNS

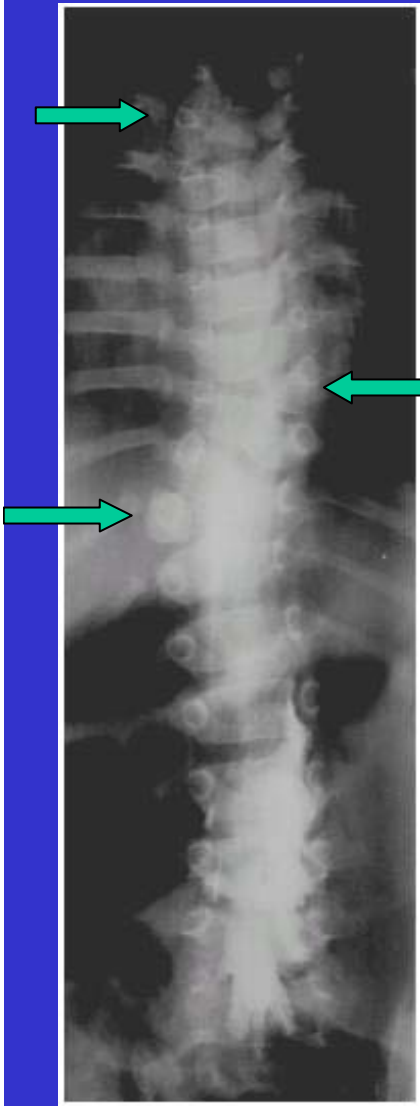
HV1/2 S

HV Trap Libre

HV Libre

Barre antéro-latérale

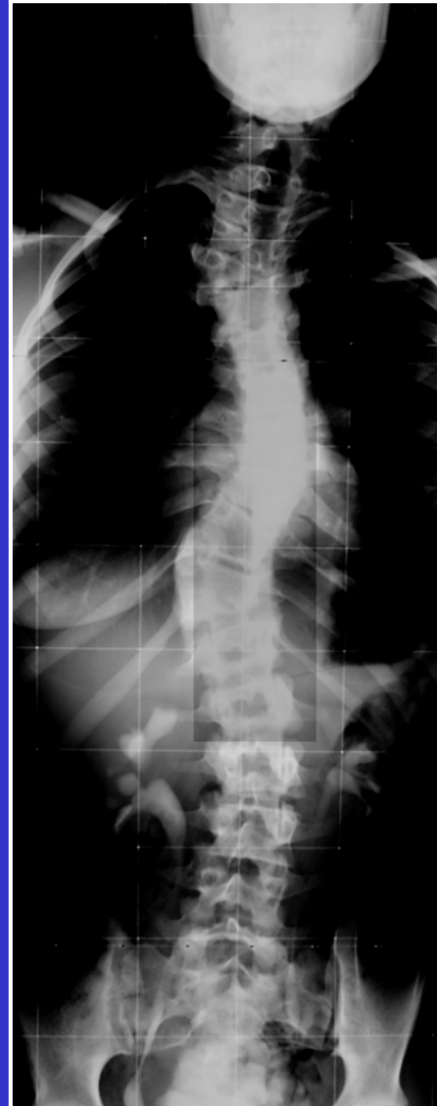
**CYPHOSE**



**8 M**



**8 A**

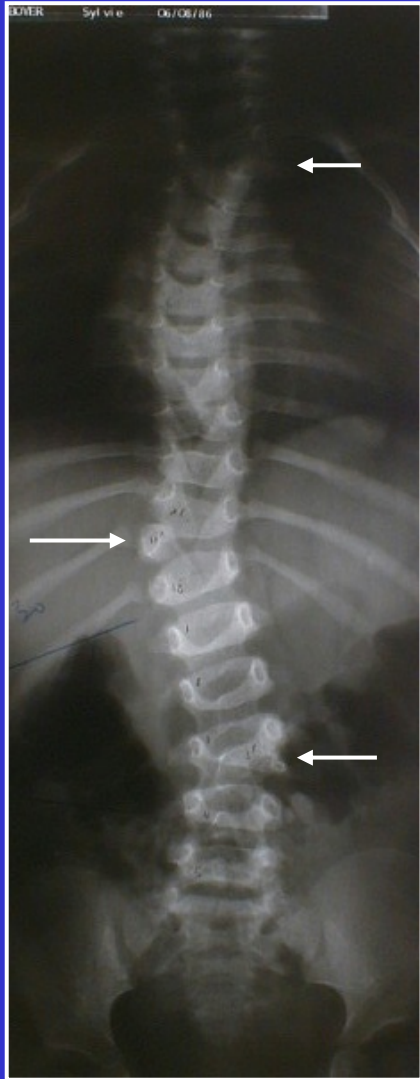


**15 A**



**15 A**





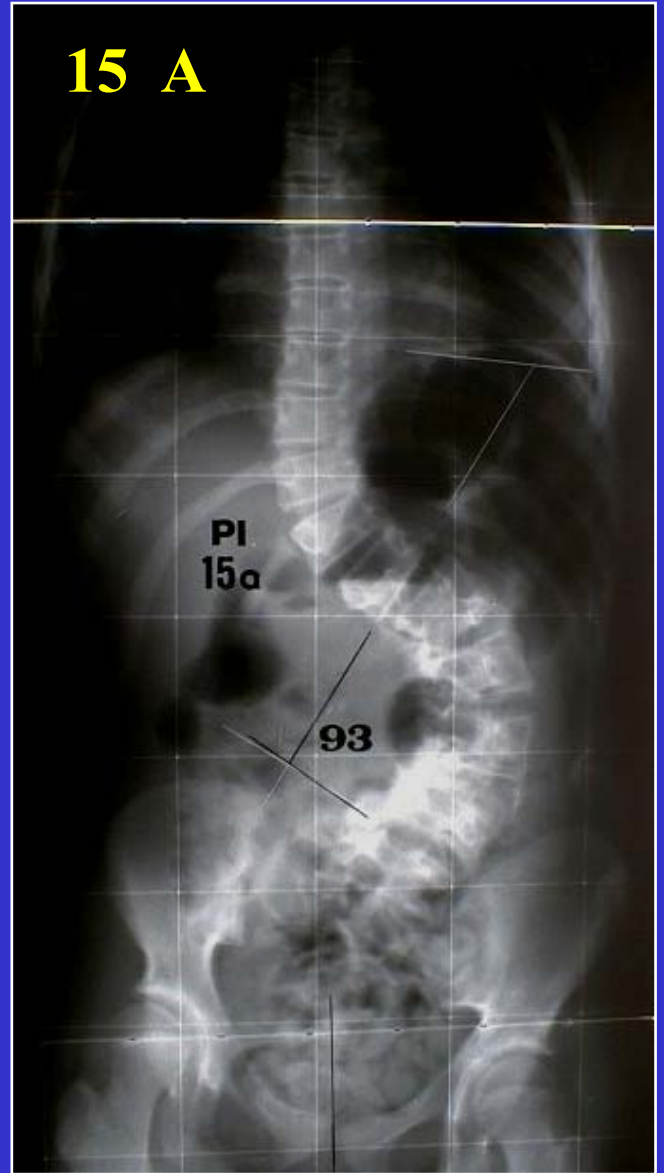
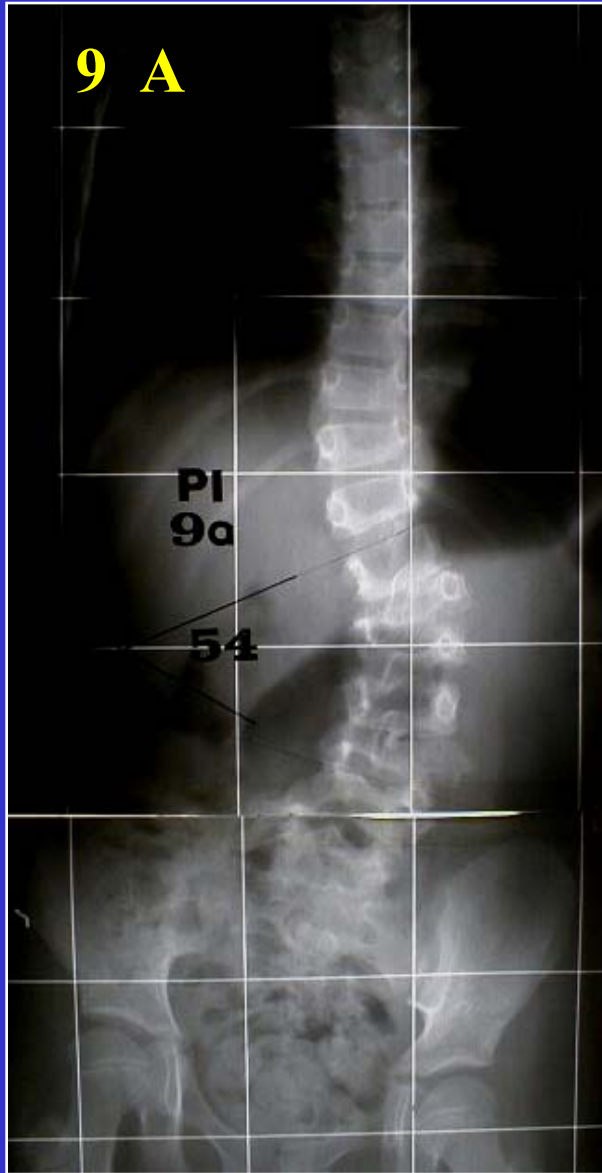
**2 A**



**10 A**



**16 A**



# CLASSIFICATION DES SCOLIOSES CONGENITALES

**SHANDS et BUNDENS ( 1956)**

**MAC EWEN Non publié**

**COTREL ( 1979)**

A.R. Shands, W.D. Bundens.

Congenital deformities of the spine. An analysis of roentgenograms of 700 children  
Bul. Hosp. Joint Dis. 1956, 17; 110-133

Y. Cotrel

Scolioses congénitales: classification et pronostic

Association Lyonnaise pour le développement des études du rachis

Bosc frères Ed. 1979; 129-140



# CLASSIFICATION DES SCOLIOSES CONGENITALES

**WINTER 1968**

- I Inclassables**
- II Fusions costales**
- III Défaut partiel unilatéral de formation (vertèbre trapézoïdale)**
- IV Absence complète unilatérale de formation (hémi vertèbre)**
- V Absence bilatérale de segmentation (fusion vertébrale)**
- VI Absence unilatérale de segmentation (barre)**

Congenital scoliosis. A study of 234 patients treated and untreated. Part I Natural history

R.B. Winter, J.H. Moe, V.E. Eilers

J Bone Joint Surg. 1968, 50 A, 1; 1-15

# CLASSIFICATION DES SCOLIOSES CONGENITALES

PAR H.V.

**NASCA 1975**

Hémivertèbre surnuméraire unique

Hémivertèbre trapézoïdale unique

Hémivertèbres multiples

Hémivertèbres multiples avec barre controlatérale

Hémivertèbres équilibrées

Hémivertèbres postérieures

Progression of congenital scoliosis due to hemivertebra R.J.  
Nasca, F.H. Stelling, H.H. Steel J Bone  
Joint Surg. 1975, 57 A, 4; 456-466

# CLASSIFICATION DES SCOLIOSES CONGENITALES

## MAC MASTER 1982

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The natural history of congenital scoliosis.  
M.J. Mc Master, K. Ohtsuka  
J Bone Joint Surg. 1982, 64 A, 8; 1128- 47

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# I INCLASSABLE



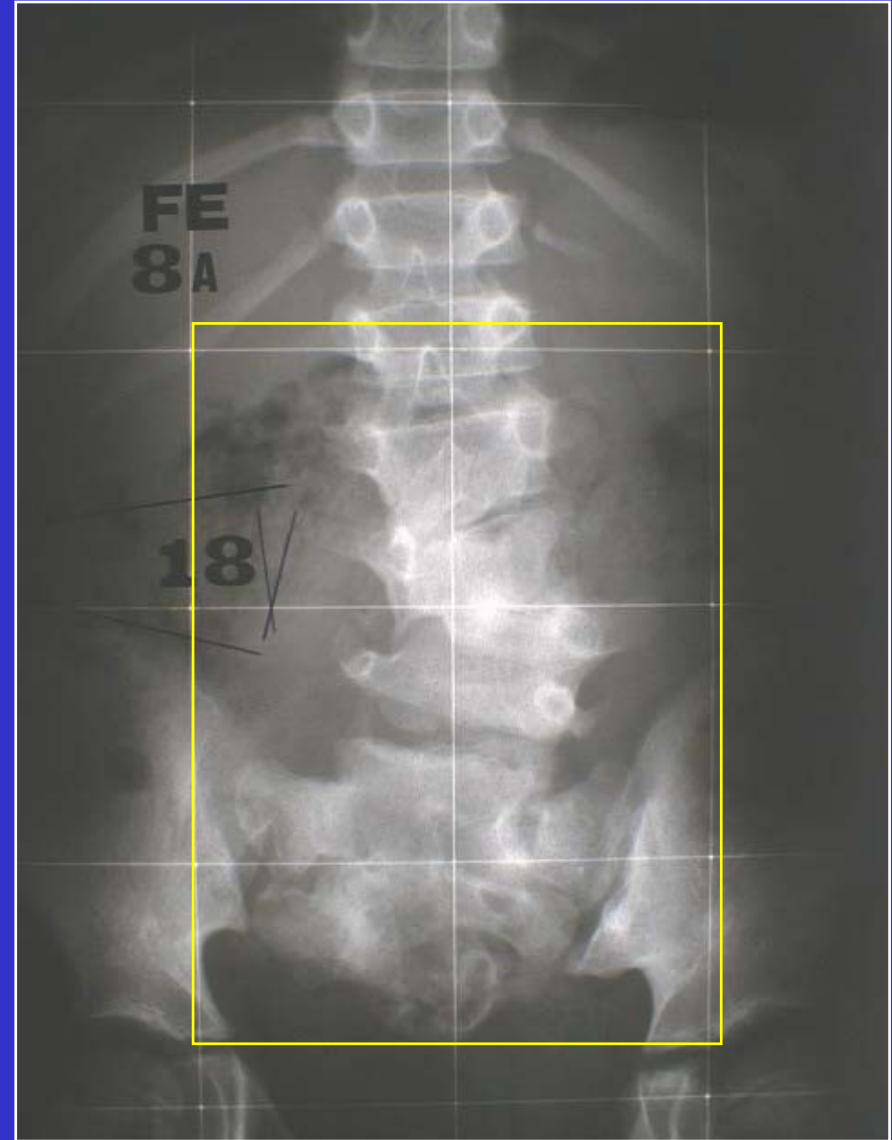
**AGENESIE COSTO-VERTEBRALE**

**Défaut de chondrification**

**NAISSANCE**



**8 A**



**DEFAUT D' OSSIFICATION**

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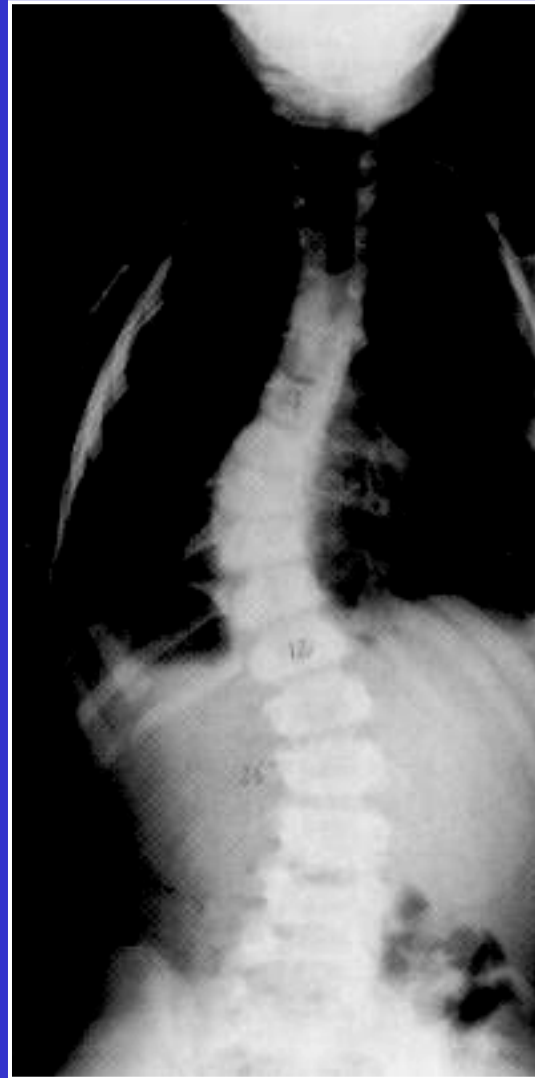
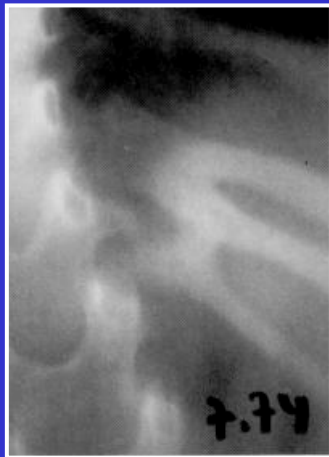
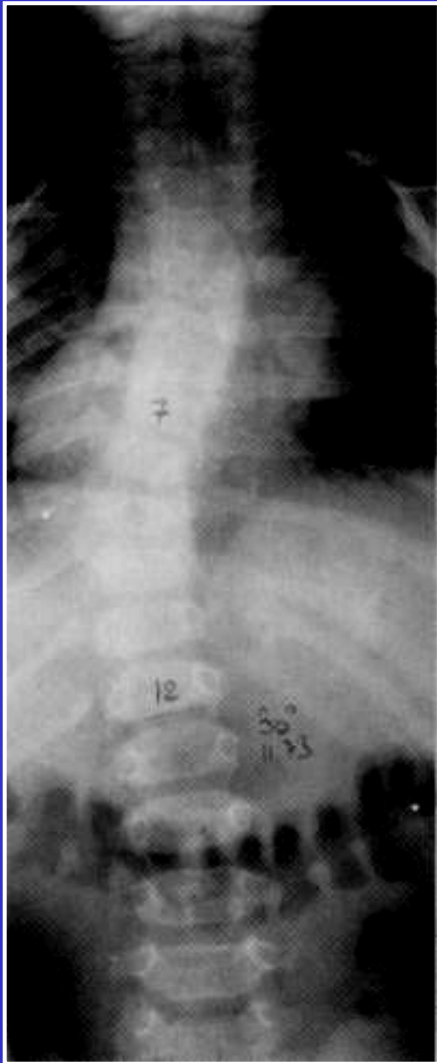
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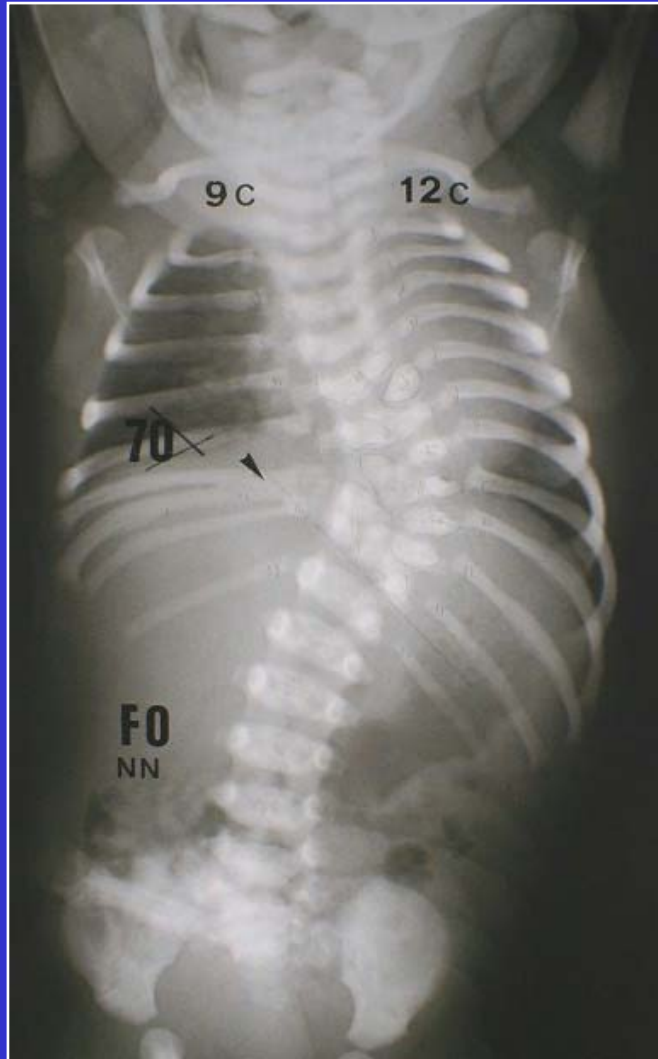
**V Défaut de segmentation**

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## II FUSION COSTALES

9 Côtes

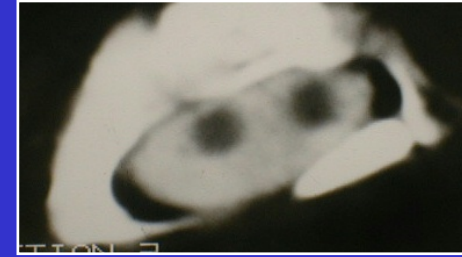


12  
Côtes



PUZZLE VERTEBRAL

## II FUSION COSTALE + BARRE



## DIASTEMATOMYELIE



# CLASSIFICATION DES SCOLIOSES CONGENITALES

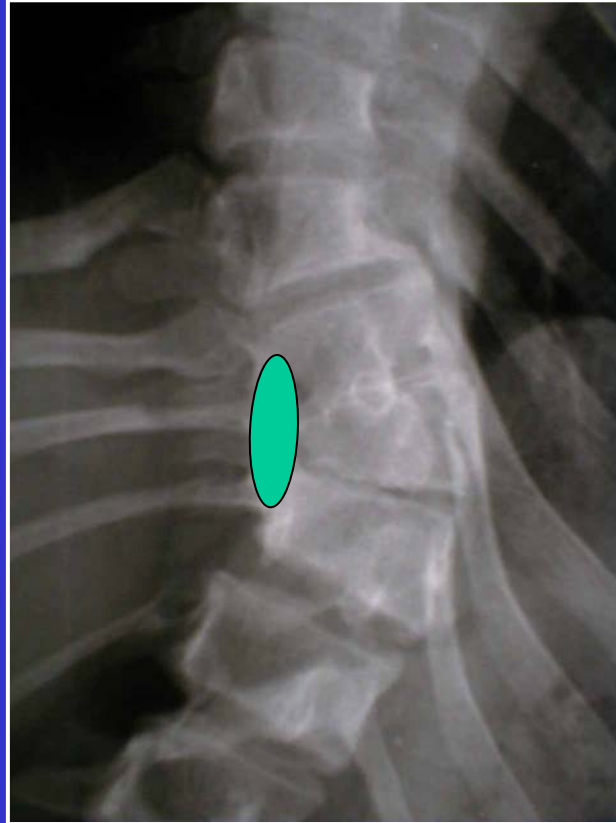
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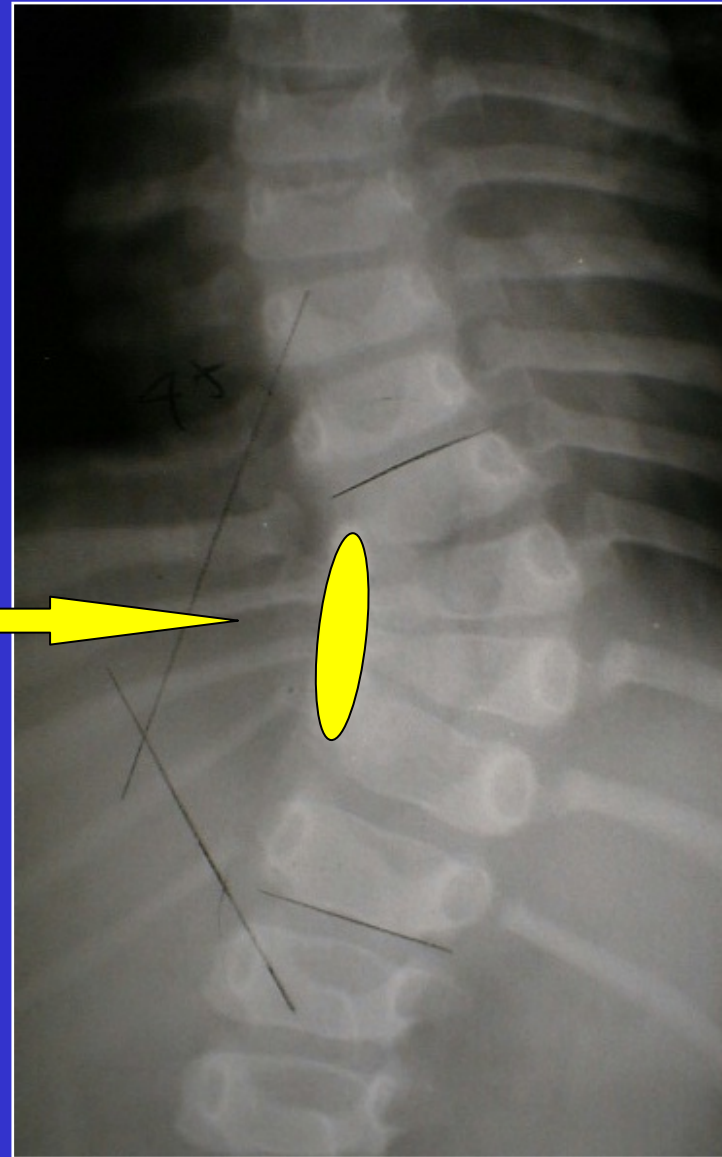
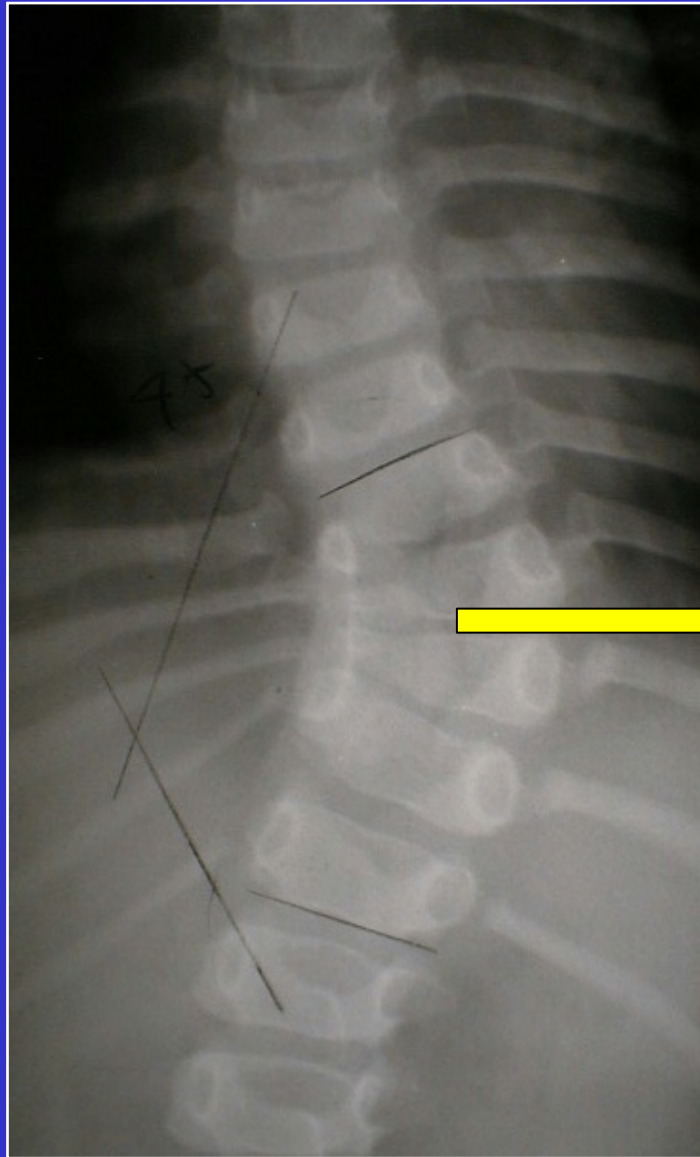
The natural history of congenital scoliosis.  
M.J. Mc Master, K. Ohtsuka  
J Bone Joint Surg. 1982, 64 A, 8; 1128- 47



# BARRE



# BARRE LATERALE



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59 patients consécutifs avec scoliose congénitale par  
**Hémivertèbre + barre controlatérale**

Dysraphisme spinal fermé dans 41% des cas

**4 Niveaux:** thoracique haut  
Thoracique moyen  
Thoracolumbaire  
Lombaire

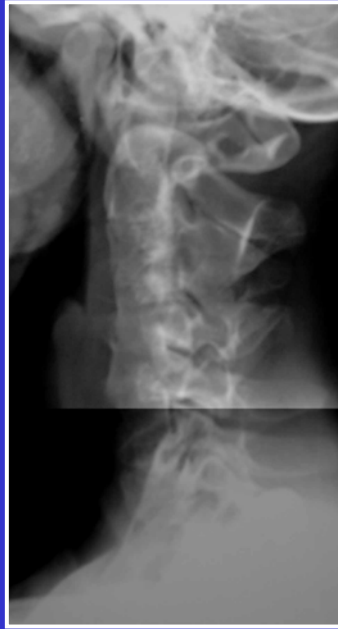
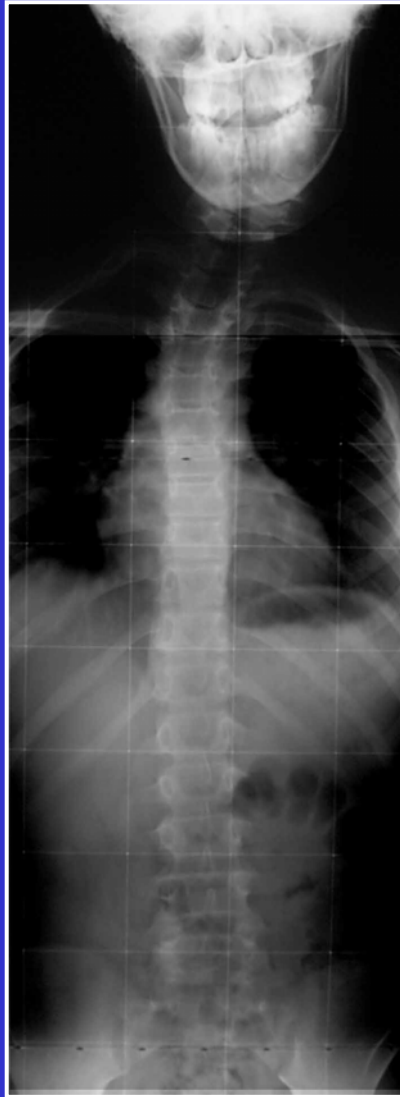


Congenital scoliosis caused by a unilateral failure of vertebral segmentation with controlateral hemivertebrae.

McMaster MJ.

Spine 1998 May 1 ; 23 (9): 998-1005

# Thoracique haut

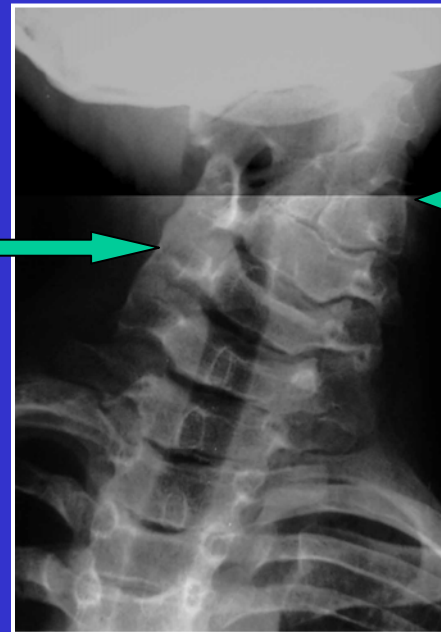


**15 A**

**BARRE**

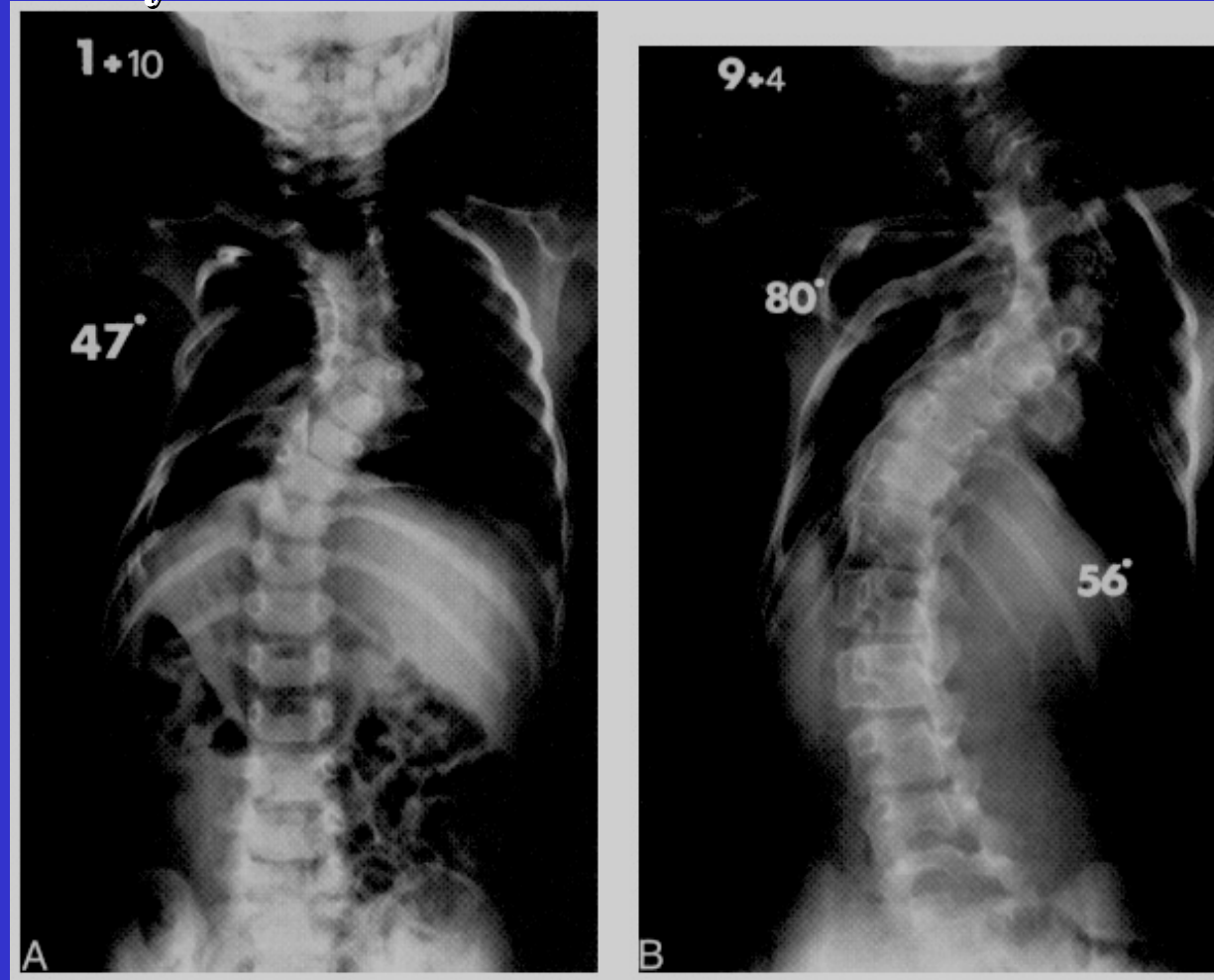


**H.V**





## Thoracique moyen

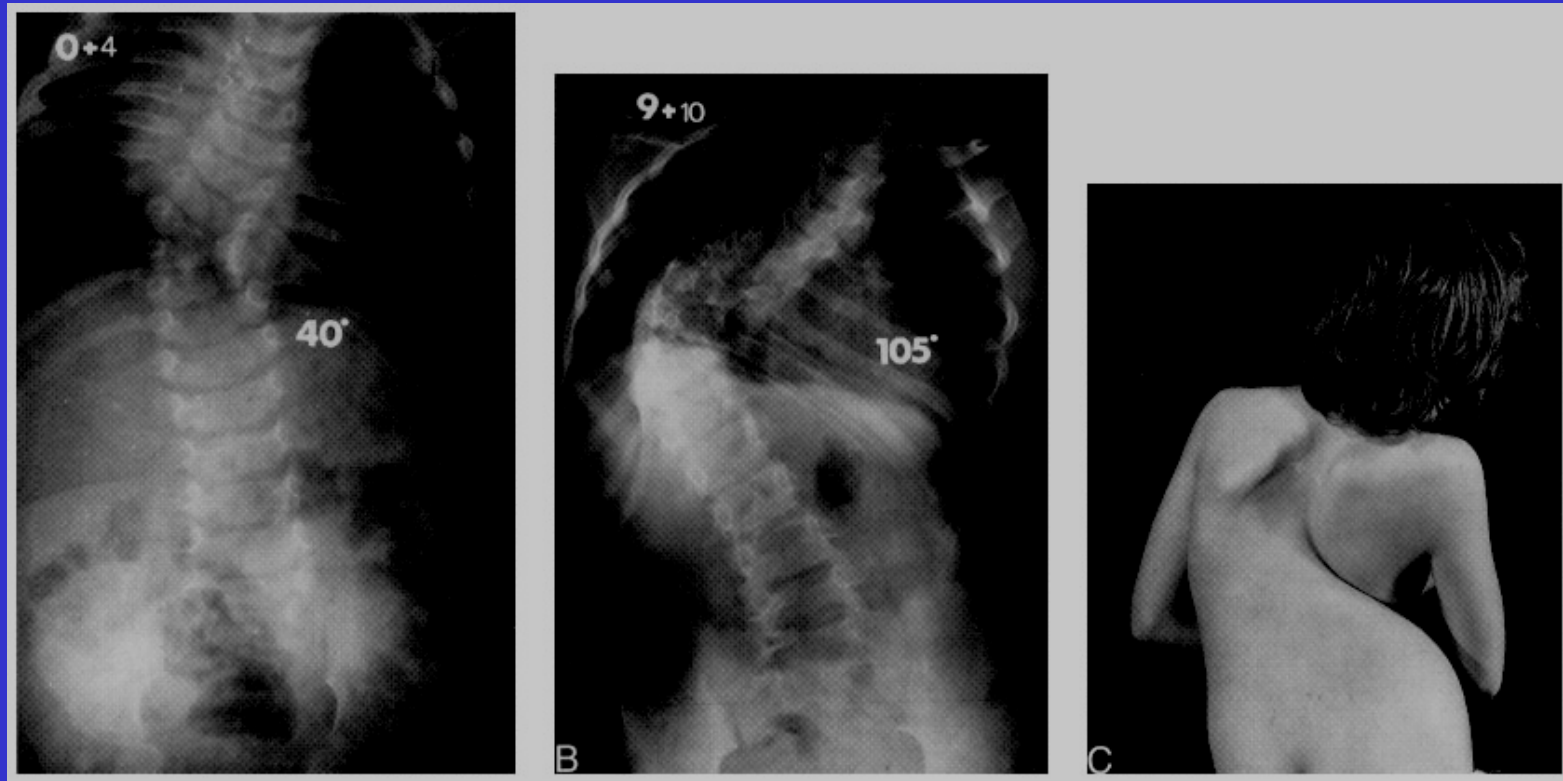


Congenital scoliosis caused by a unilateral failure of vertebral segmentation with contralateral hemivertebrae.

McMaster MJ.

Spine 1998 May 1 ; 23 (9): 998-1005

## Thoraco lombaire

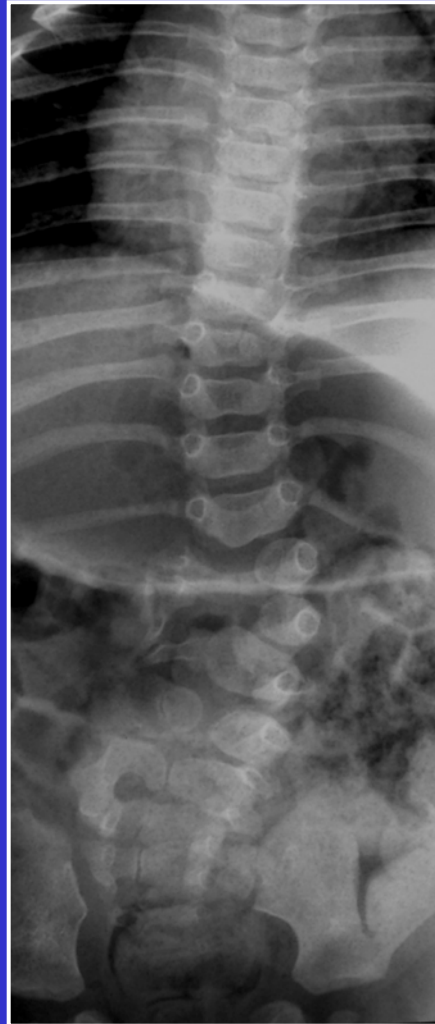


Congenital scoliosis caused by a unilateral failure of vertebral segmentation with contralateral hemivertebrae.

McMaster MJ.

Spine 1998 May 1 ; 23 (9): 998-1005

# Lombaire



4.2003

7 M

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# Hémivertèbres et Scolioses

Type d'hémivertèbre

Localisation de l'hémivertèbre

Nombre d'hémivertèbres et leurs positions respectives

Age du patient



# Hémivertèbres et Scolioses

Type d'hémivertèbre

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Nombre d'hémivertèbres et leurs positions respectives

Age du patient

## **HEMIVERTEBRE COMPLETEMENT SEGMENTEE**

encore appelée **HEMIVERTEBRE LIBRE**

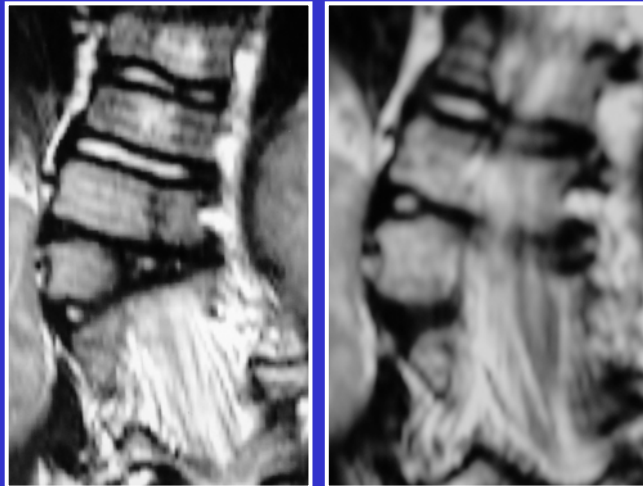


La plupart du temps la supposée hémivertèbre libre est complètement segmentée en avant, séparée des corps vertébraux adjacents par des structures de croissance mais, en arrière, cette même hémivertèbre présente une hémilame en fusion avec la lame de la vertèbre sus ou sous jacente.

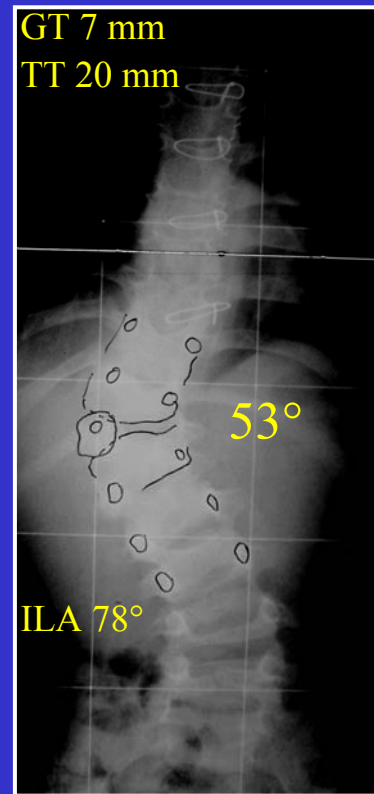
Ce point est important à connaître lors de la résection d'une hémivertèbre.

TRE. 15.2.95

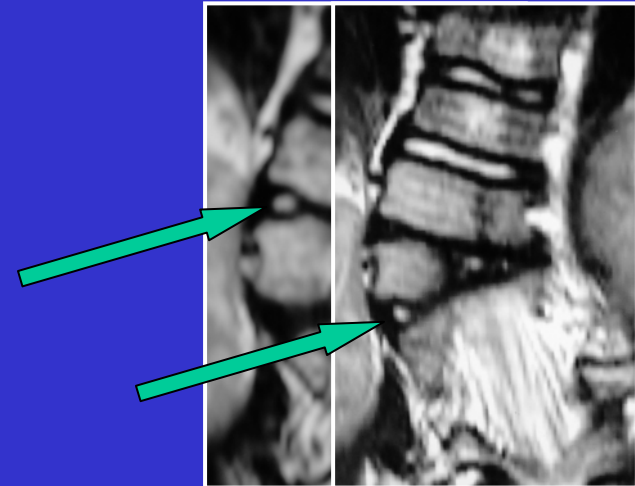
## H.V. LIBRE T 12 L 1 DROITE



L'IRM est très utile pour classier l'H.V.



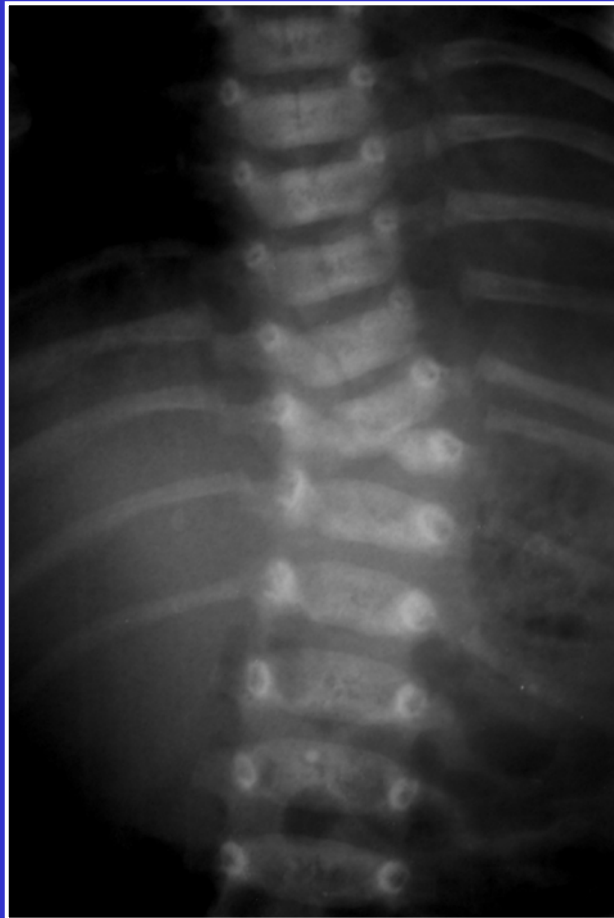
4 A 2 M



**IRM pondération T2**

**Signal de nucléus  
au dessus et en  
dessous de l'H.V.**

# HEMIVERTEBRE SEMI SEGMENTEE

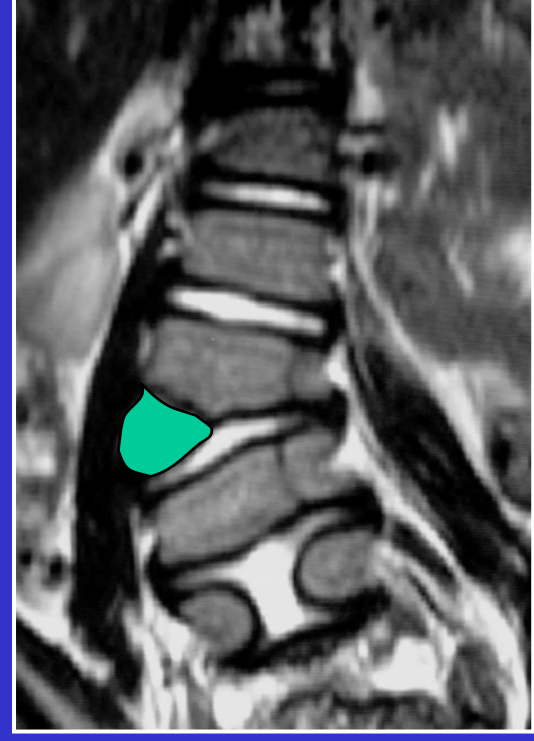
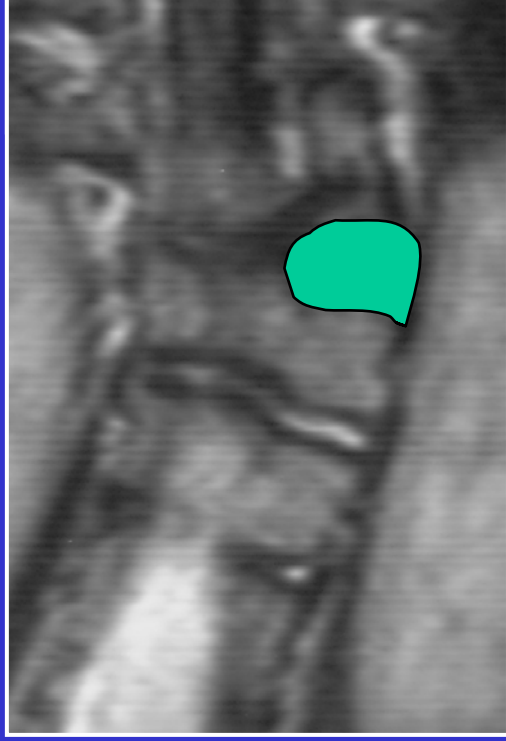
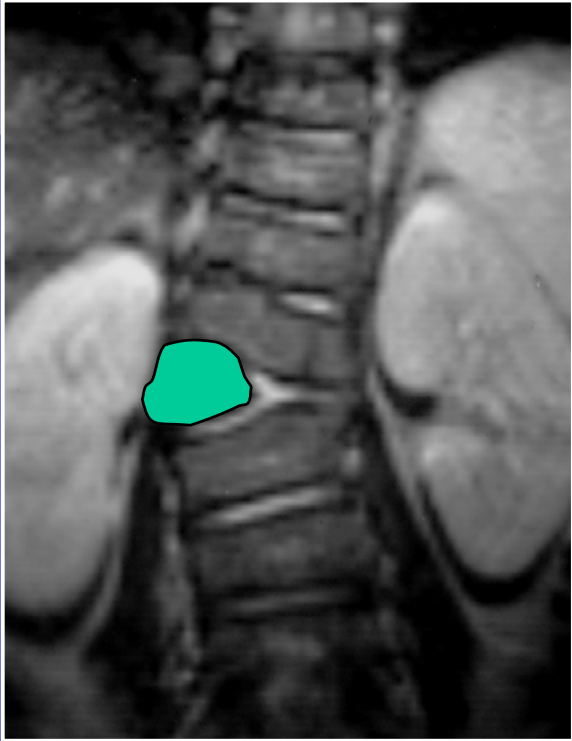
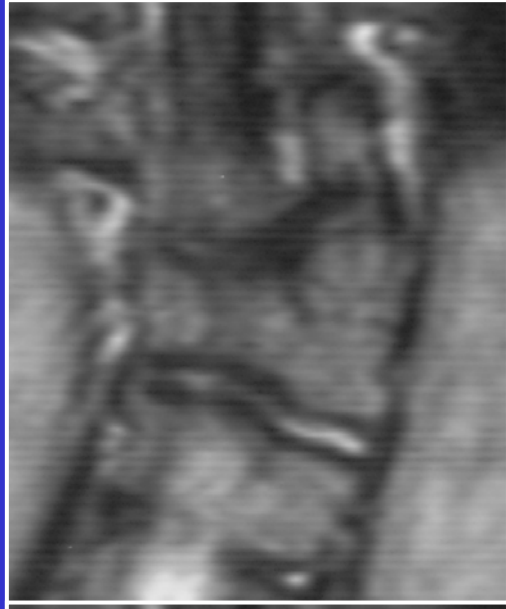


**Naissance**

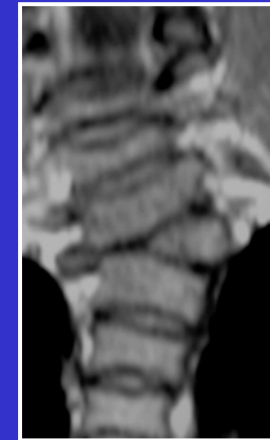
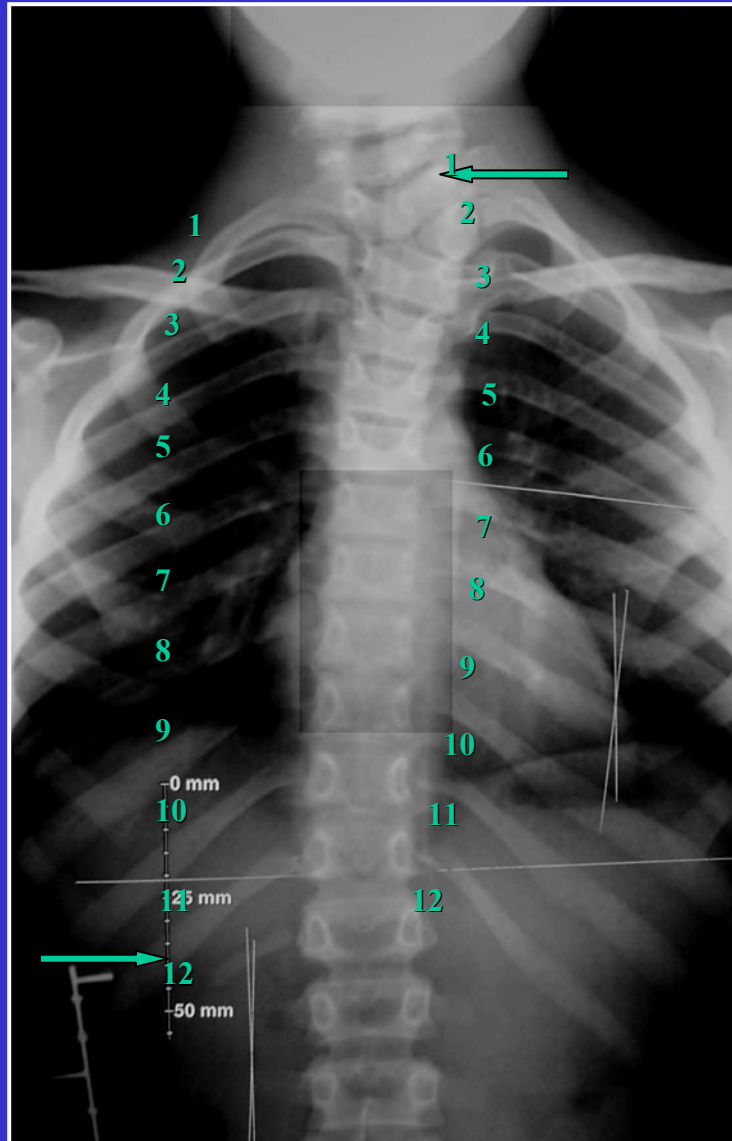
ou en HEMI FUSION



**8 M**



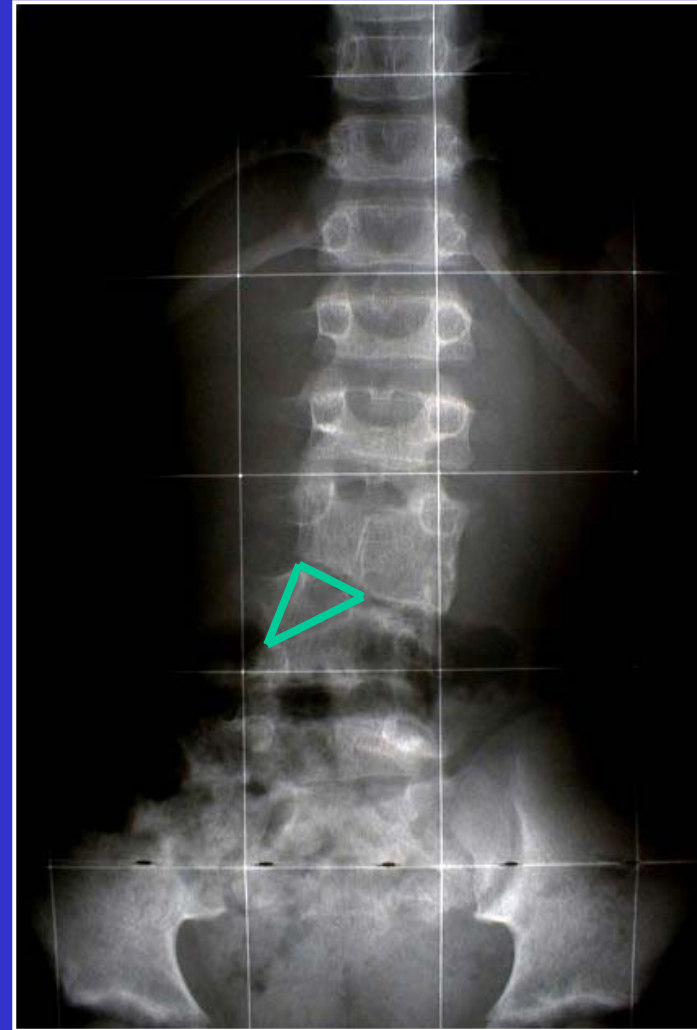
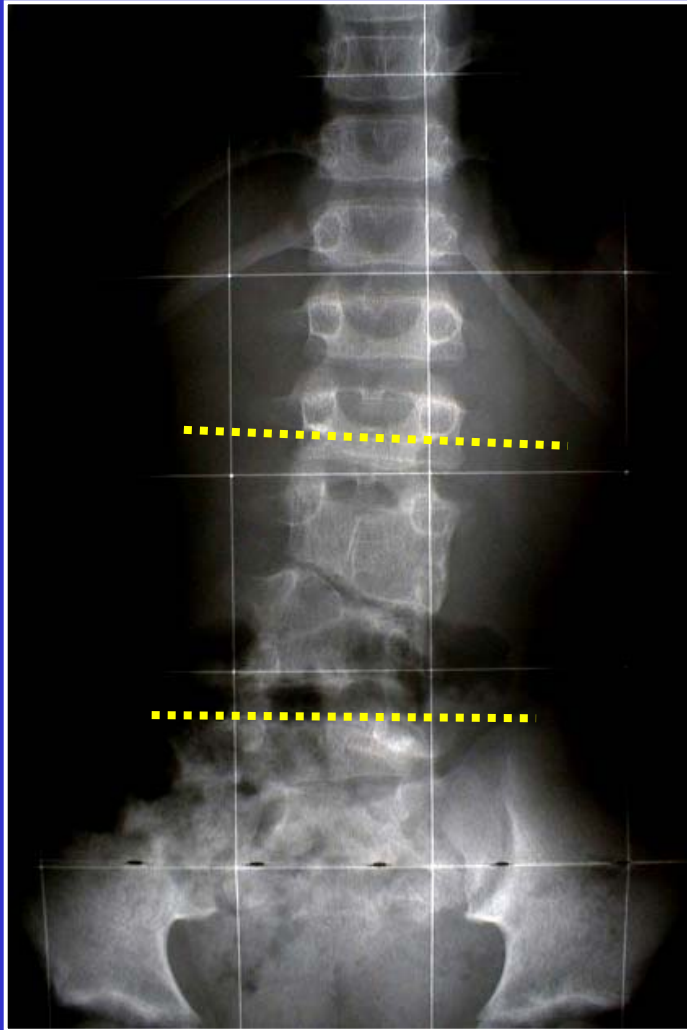




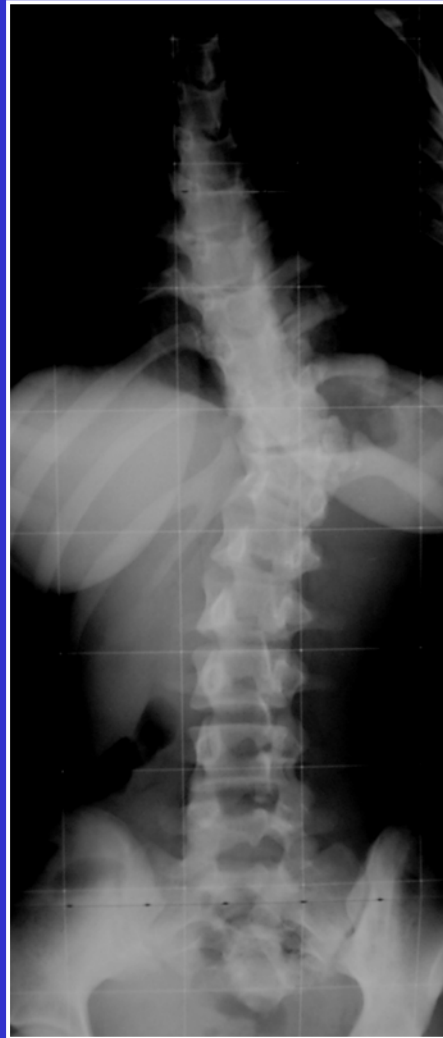
**10 A**

**HEMIVERTEBRE NON SEGMENTEE**

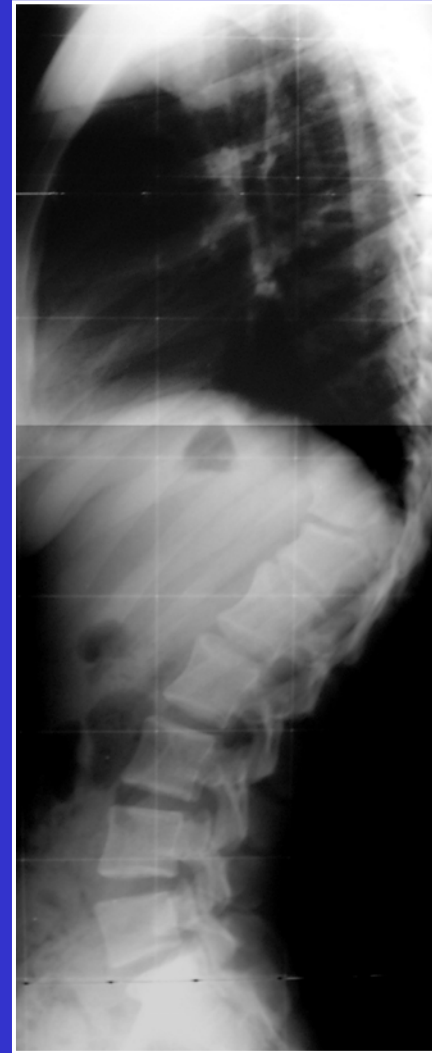
# HEMIVERTEBRE INCARCEREE

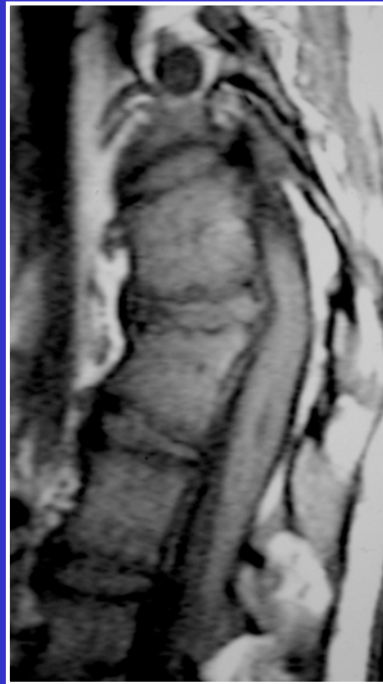
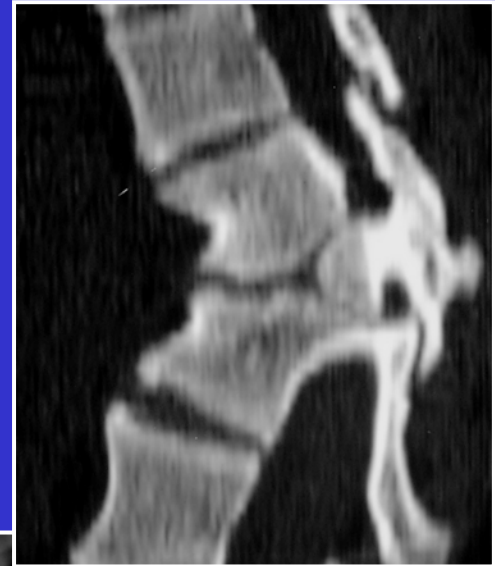
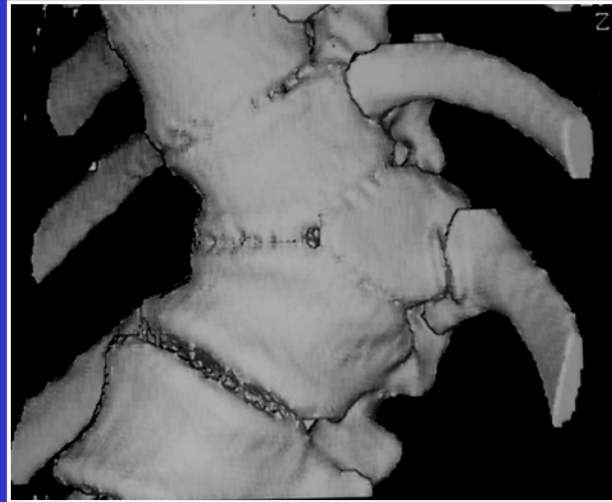


# HEMIVERTEBRE « POSTERIEURE »

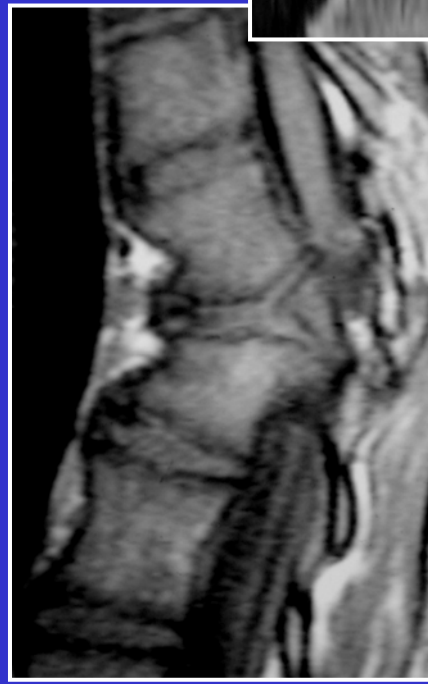


12 A





12 Y



# Hémivertèbres et Scolioses

Type d'hémivertèbre

**Localisation de l'hémivertèbre**

Nombre d'hémivertèbres et leurs positions respectives

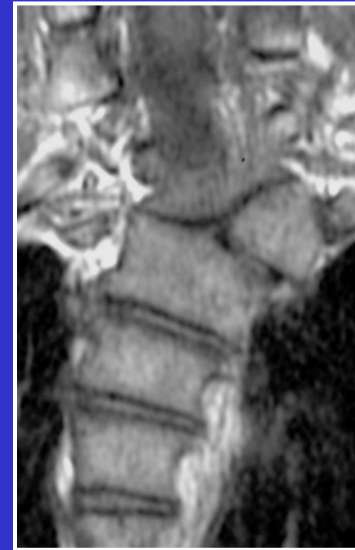
Age du patient



# CERVICO - THORACIQUE

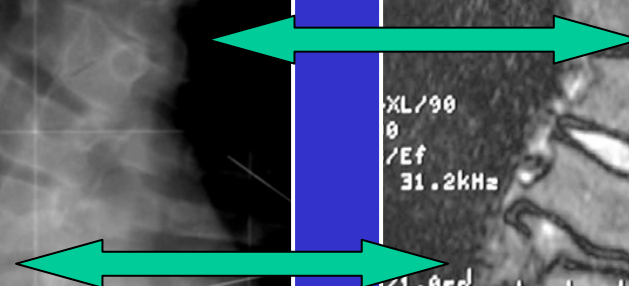
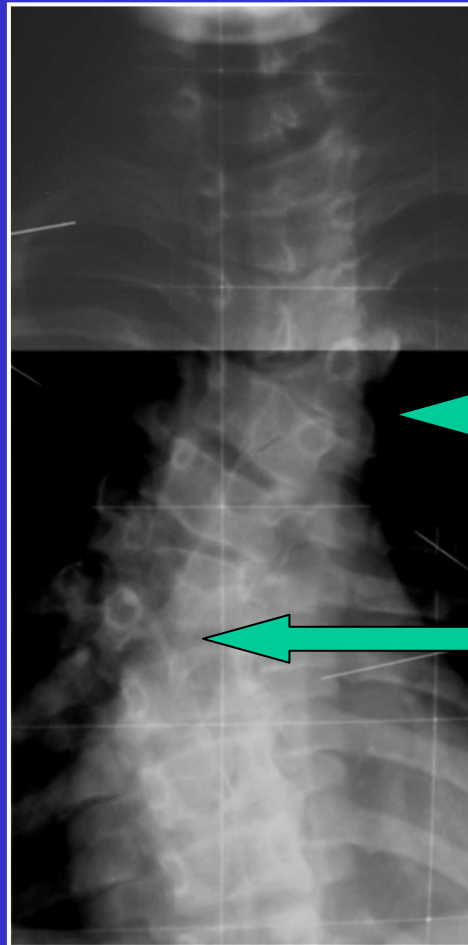
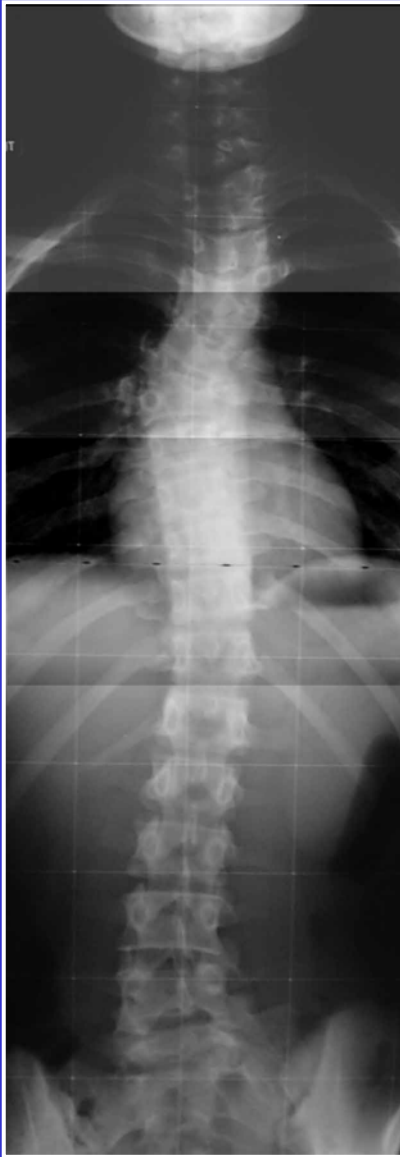


**16 A**

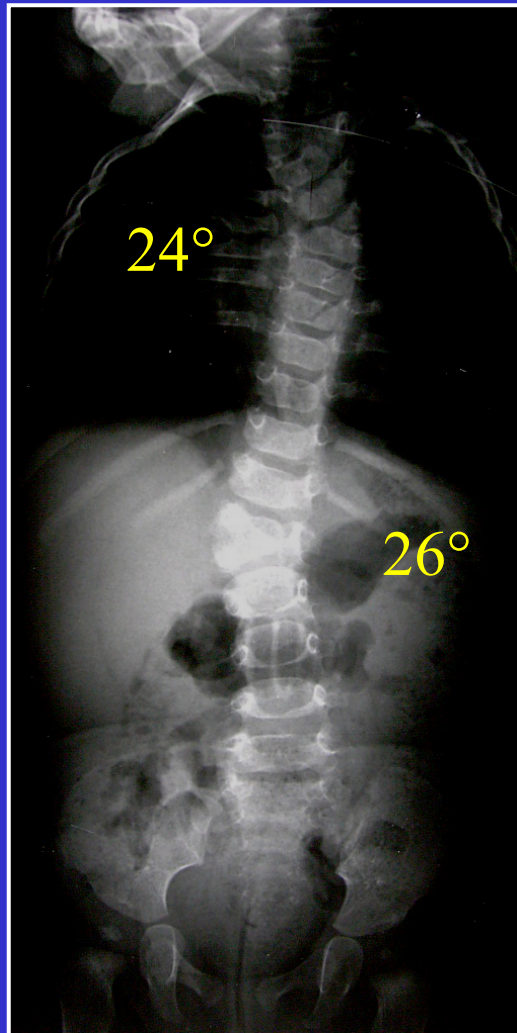


# THORACIQUE

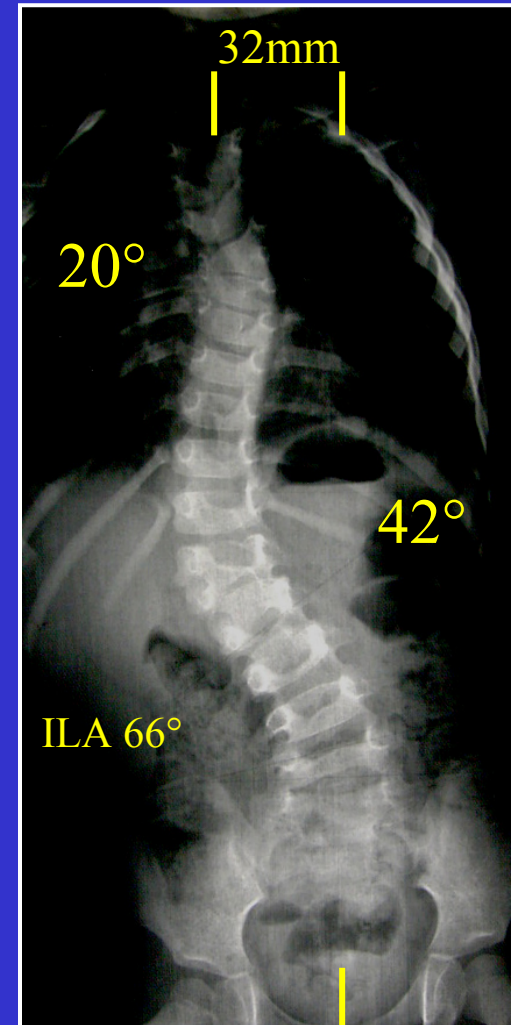
14 A



# THORACO - LOMBAIRE

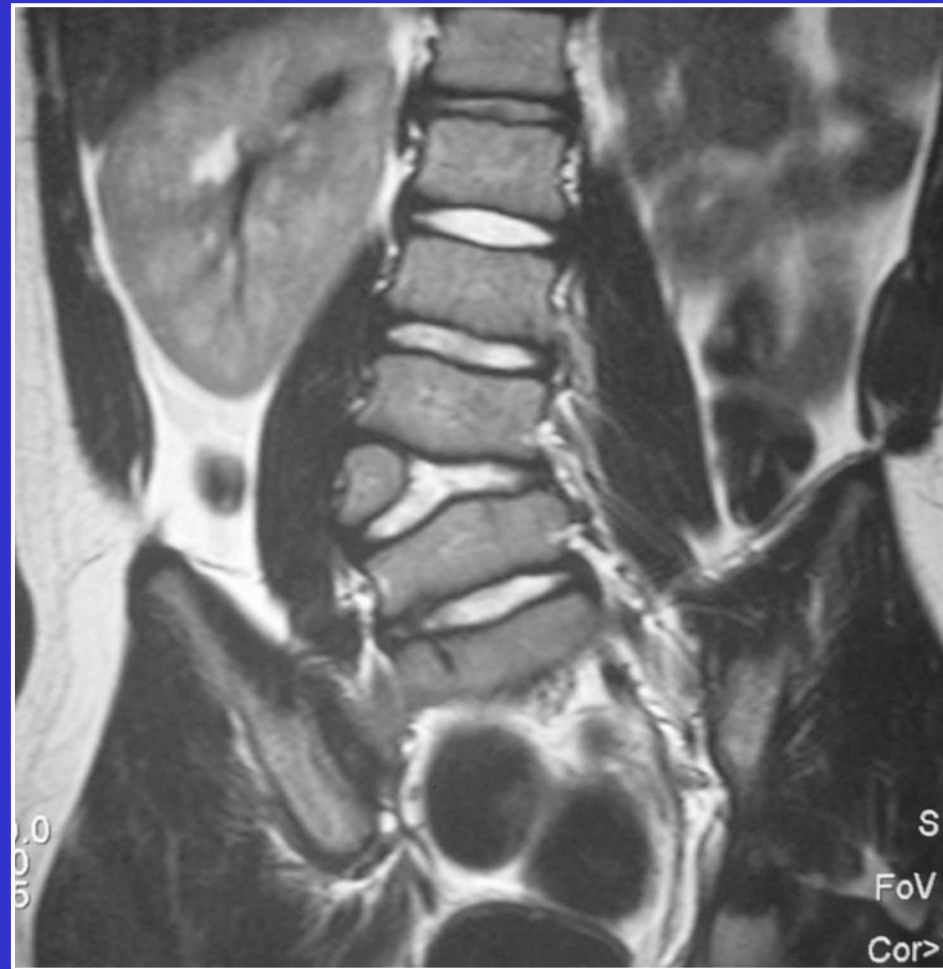


**7 Mois**



**18 Mois**

# LOMBAIRE



3. 2002

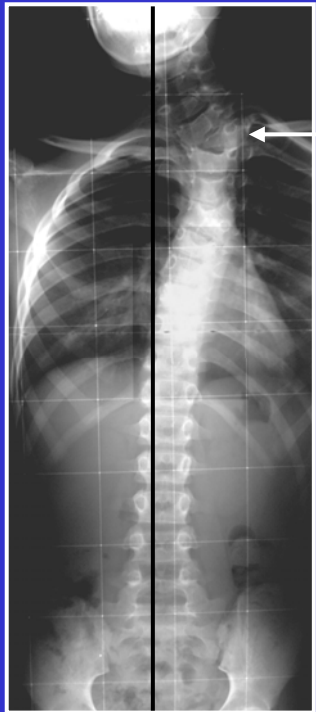
# LOMBO - SACREE



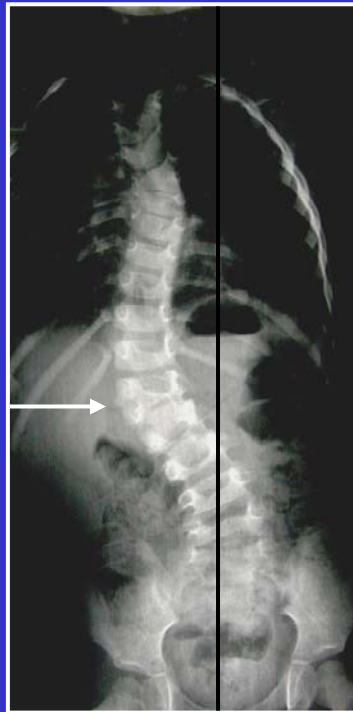
5 A 11 M



## HEMIVERTEBRE « JONCTIONNELLE »



Cervico-thoracique



Thoraco-lombaire



Lombo-sacrée

TRANSLATION DU TRONC



# Hémivertèbres et Scolioses

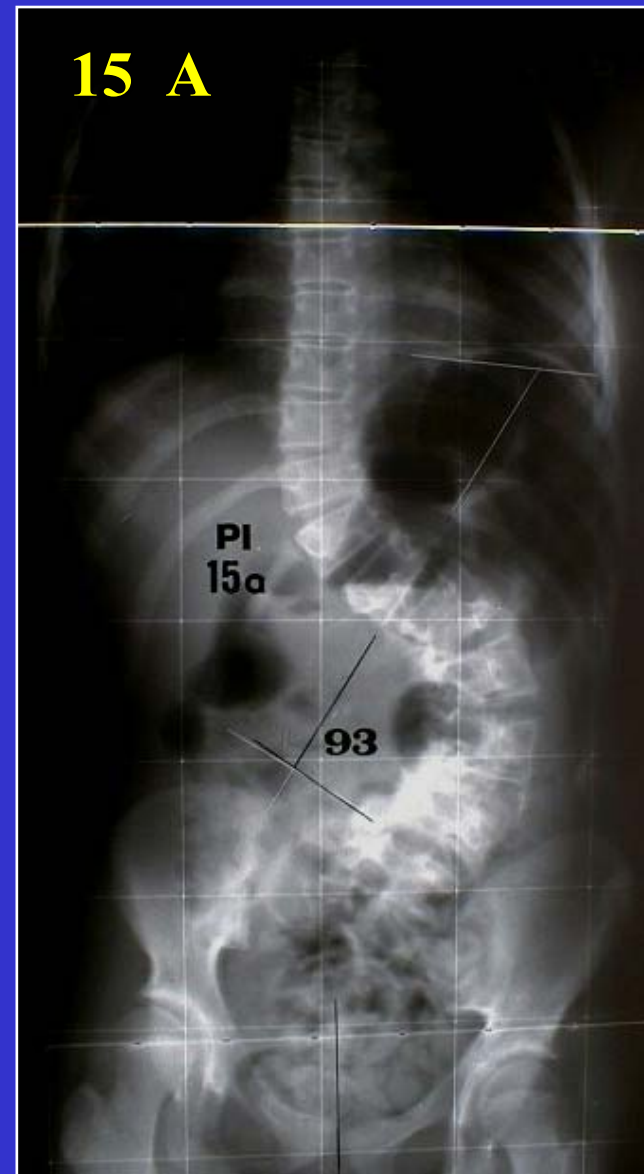
Type d'hémivertèbre

Localisation de l'hémivertèbre

**Nombre d'hémivertèbres et leurs positions respectives**

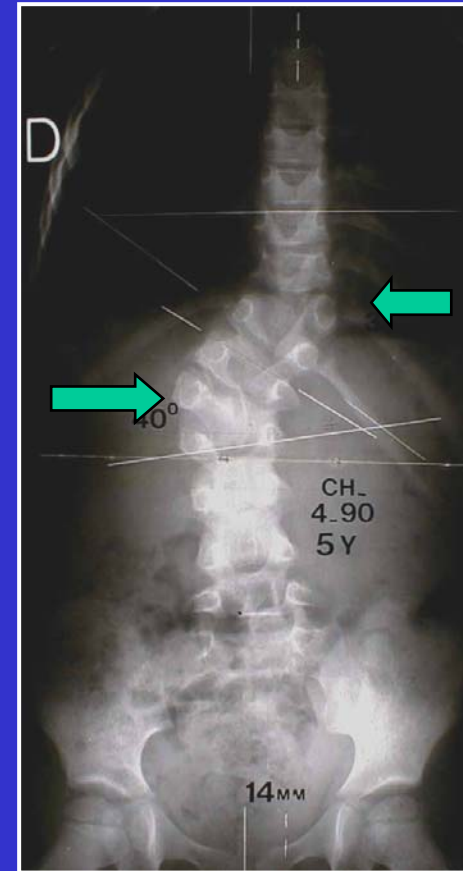
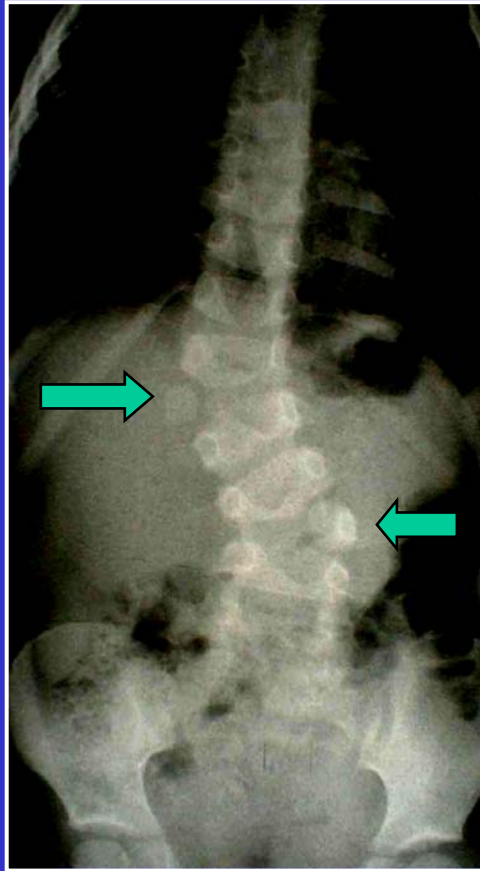
Age du patient

## H.V. IPSI LATERALES



## H.V. ALTERNEES

S.B. Shawen, P.J. Belmont, T.R. Kuklo, B.D. Owens, K.F. Taylor, R. Kruse, D.W. Polly Hemimetameric segmental shift: a case series and review. Spine 2002 Dec 15,27 (24): E 539 - 44



« La correction chirurgicale est plus généralement proposée lorsque l'H.V. caudale est située entre les jonctions thoraco-lombaire et lombo-sacrée. »

# Hémivertèbres et Scolioses

Type d'hémivertèbre

Localisation de l'hémivertèbre

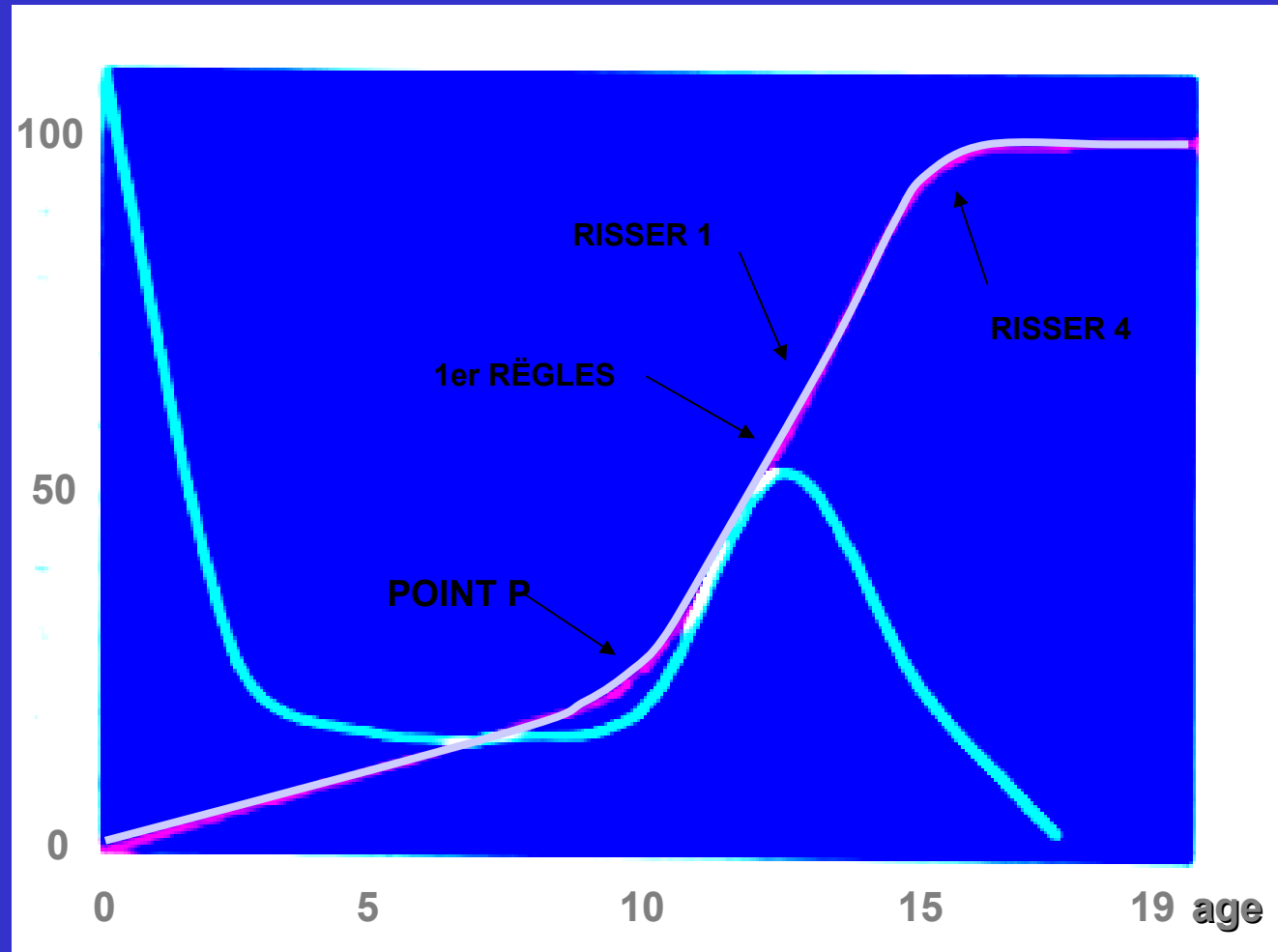
Nombre d'hémivertèbres et leurs relations

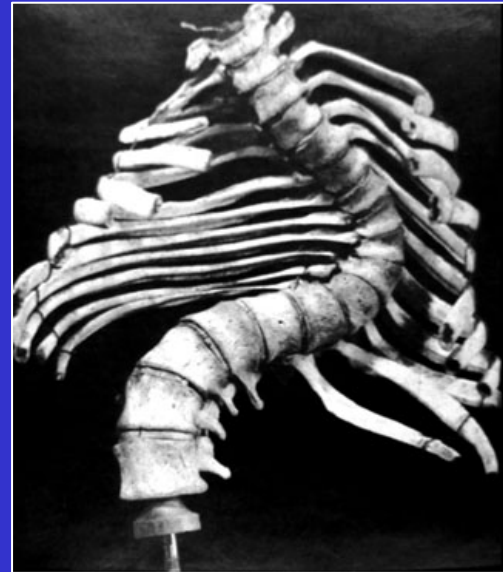
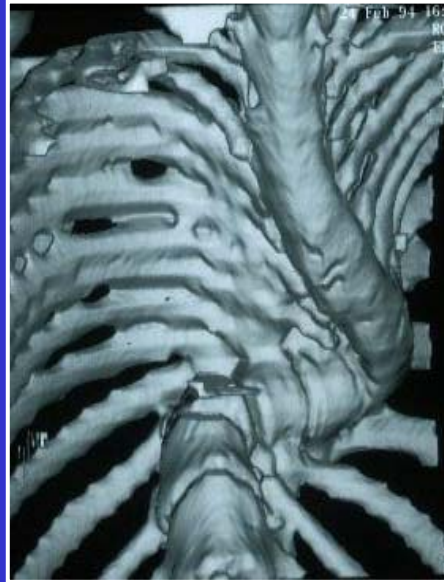
**Age du patient**

# COURBE DE Mme DUVAL - BEAUPERE

—  
**Angle de Cobb**

—  
**Vitesse de  
croissance  
rachidienne**





## **DISLOCATION ROTATOIRE**

**J. DUBOUSSET**



# **OPTIONS THERAPEUTIQUES**

## **NON CHIRURGICALES**

**Le corset est considéré comme sans effet sur le contrôle de l'évolutivité des scolioses congénitales.....**

**Mais effet sur les contre courbures ?**

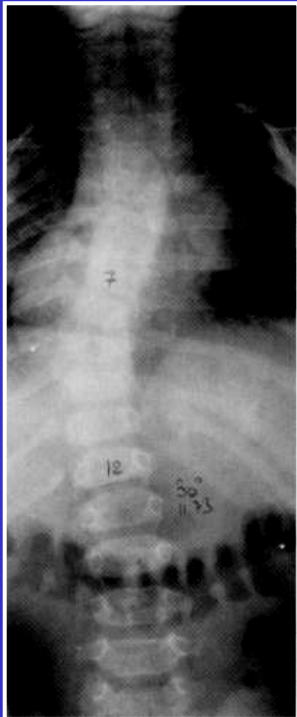
## OPTIONS THERAPEUTIQUES

### Chirurgie « costale »

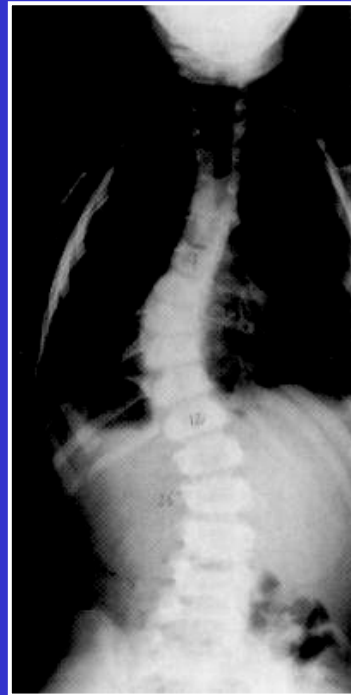
Scoliosis and fused ribs. A case report Damsin JP, Cazeau C, Carlioz H.  
Spine 1997 May 1; 22 (9): 1030-2

R.M. Campbell, M.D. Smith, T.C. Mayes, J.A. Mangos, D.B. Willey-Courand, N. Kose, R.F. Pinero, M.E. Alder, H.L. Duong, J.L. Surber. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis.  
J Bone Joint Surg. 2003, 85 A (3) ; 399 - 408

J.F. Taylor, R. Roaf, R. Owen, G. Bentley, R. Calver, R.S. Jones, M. Thorneloe  
Costodesis and contralateral rib release in the management of progressive scoliosis.  
Acta orthop. Scand. 1983, 54; 603 - 12



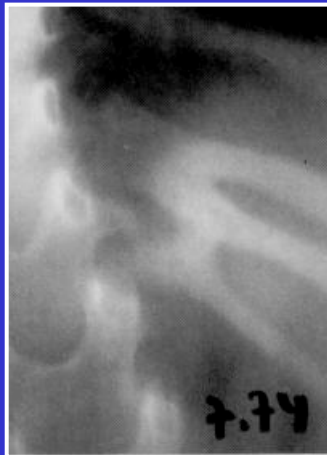
*8 Mois*



*4 A 3 M*



*18 A*



*4 A 3 M*

Scoliosis and fused ribs. A case report  
Damsin JP, Cazeau C, Carlioz H.  
Spine 1997 May 1; 22 (9): 1030-2

## THE EFFECT OF OPENING WEDGE THORACOSTOMY ON THORACIC INSUFFICIENCY SYNDROME ASSOCIATED WITH FUSED RIBS AND CONGENITAL SCOLIOSIS

CAMPBELL, ROBERT M. JR. MD; SMITH, MELVIN D. MD; MAYES, THOMAS C. MD; MANGOS, JOHN A. MD; WILLEY-COURAND, DONNA B. MD; KOSE, NUSRET MD; PINERO, RICARDO F. MD; ALDER, MARDEN E. DDS; DUONG, HOA L. MD; SURBER, JENNIFER L. BS

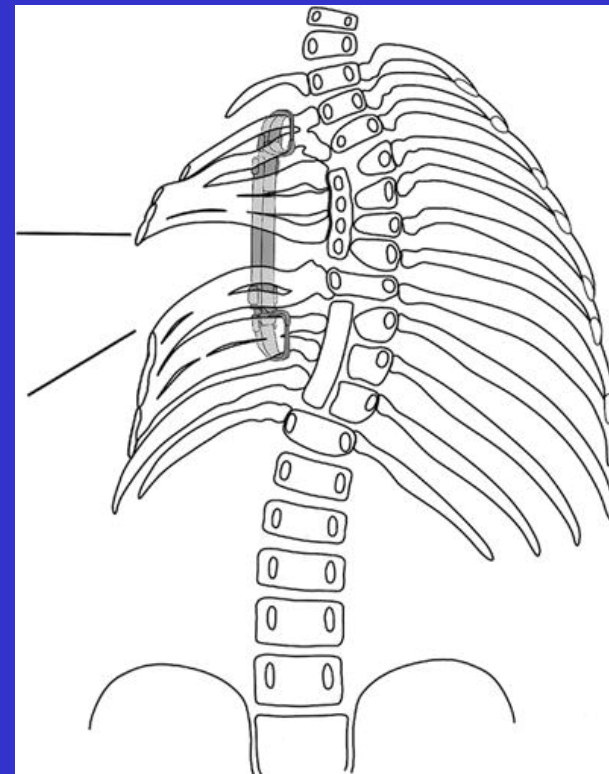
*Journal of Bone and Joint Surgery*

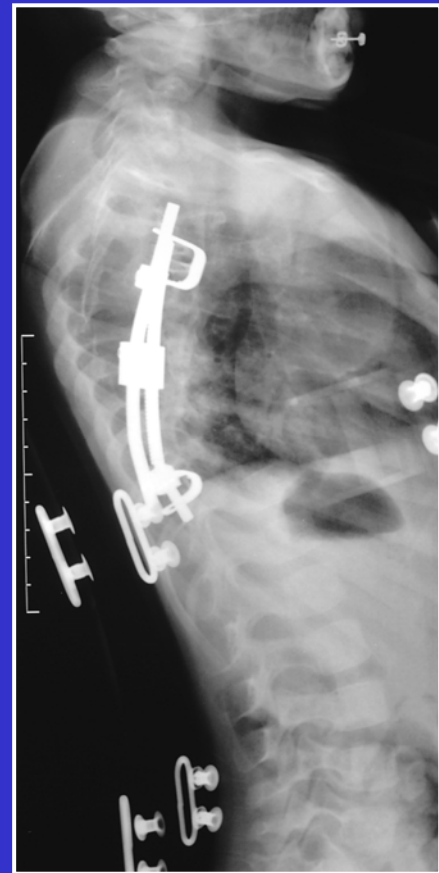
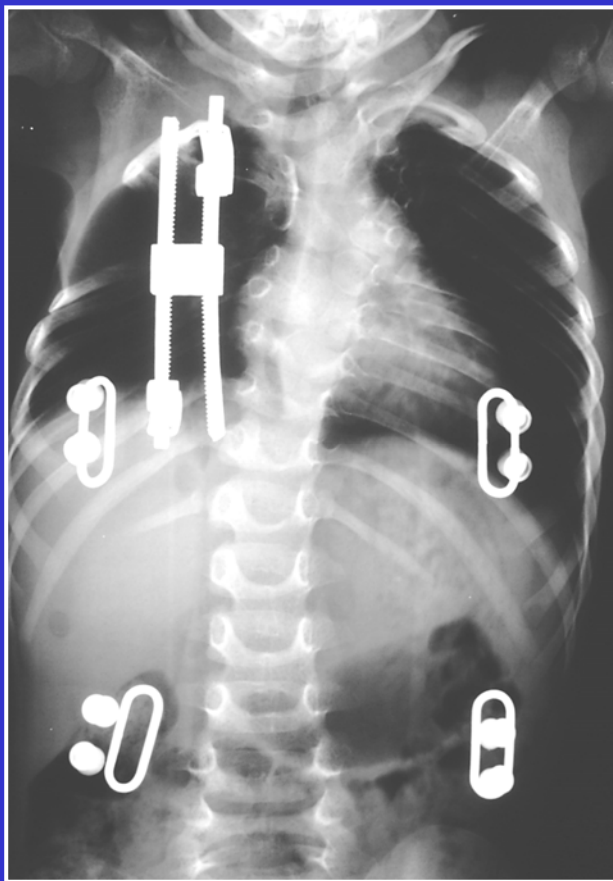
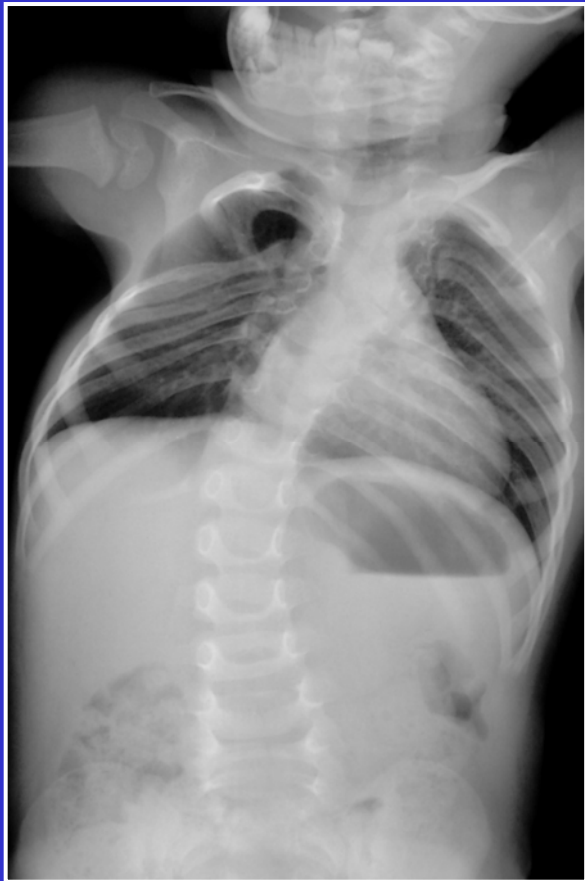
Volume 86-A(8)

August 2004

p 1659–1674

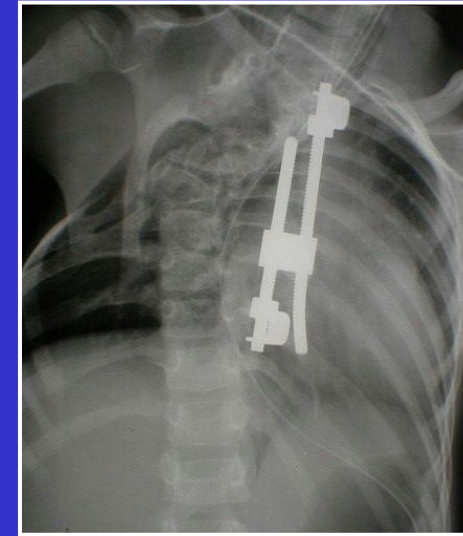
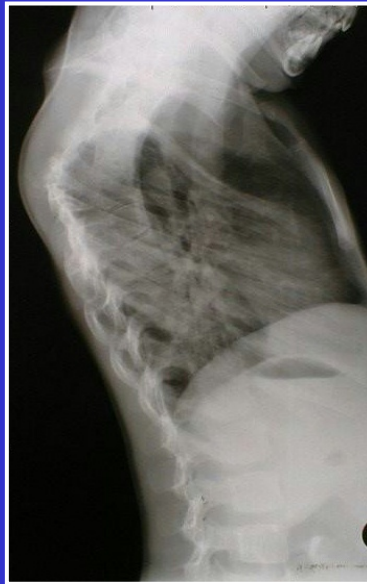
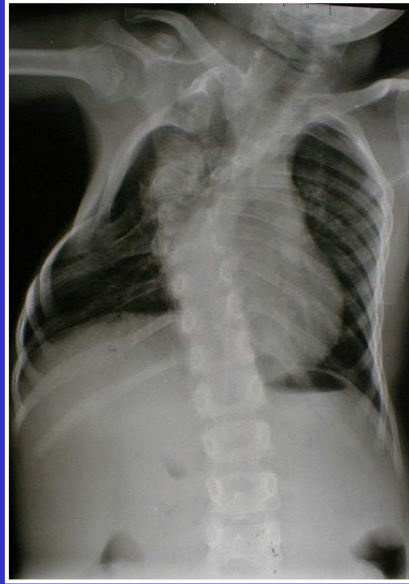
Twenty-seven patients with congenital scoliosis associated with fused ribs of the concave hemithorax had an opening wedge thoracostomy with primary longitudinal lengthening with use of a chest-wall distractor known as a *Vertical, Expandable Prosthetic Titanium Rib*.  
VEPTR





9.2002 2 A 8M







## OPTIONS THERAPEUTIQUES

Chirurgie « costale »

**Epiphysiodèse Convexe**

Convex growth arrest  
Convex hemiarthodesis  
epiphyseal arrest

R.B. Winter . Convex anterior and posterior hemiarthrodesis and hemiepiphysiodesis in young children with progressive congenital scoliosis  
J Pediatr. Orthop. 1981, 1; 361 - 66

T. Andrew, H. Piggott. Growth arrest for progressive scoliosis.  
J Bone Joint Surg. 1985, 67B ,2. 193 - 7

M. Onimus, P. Manzone, F. Michel, J.M. Chirpaz – Cerbat.  
Early operation in congenital scoliosis  
J Pediatr. Orthop. 1993 Part B, 1 . 119 - 22

## OPTIONS THERAPEUTIQUES

Chirurgie « costale »

### Epiphysiodèse Convexe

J. Dubousset, E. Katti, R. Seringe. Epiphysiodesis of the spine in young children for congenital spinal deformations. J Pediatr. Orthop. 1993 Part B, 1 . 123- 30

D.S. Marks, S.R.E. Sayampanathan, A.G. Thompson, H. Piggott.  
Long term result of convex epiphysiodesis for congenital scoliosis  
Eur. Spine J 1995, 4: 296 - 301

R.B. Winter, J.E. Lonnstein, F. Denis, H Sta-Ana de la Rosa.  
Convex growth arrest for progressive congenital scoliosis due to hemivertebrae.  
J Pediatr. Orthop. 1988, 8 (6) ; 633 - 8

# OPTIONS THERAPEUTIQUES

Chirurgie « costale »

## Epiphysiodèse Convexe

Résultat

**Pas d'effet**

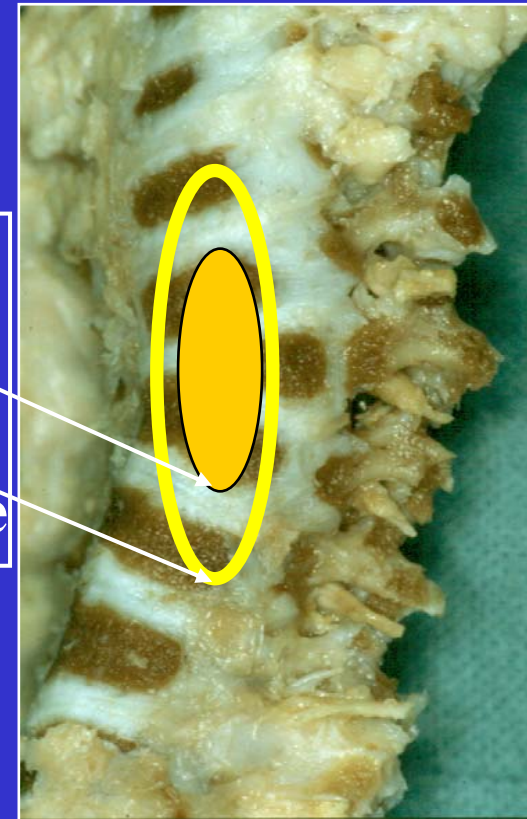
Angle de Cobb augmente

**Effet de fusion**

Angle de Cobb inchangé

**Effet d'epiphysiodèse**

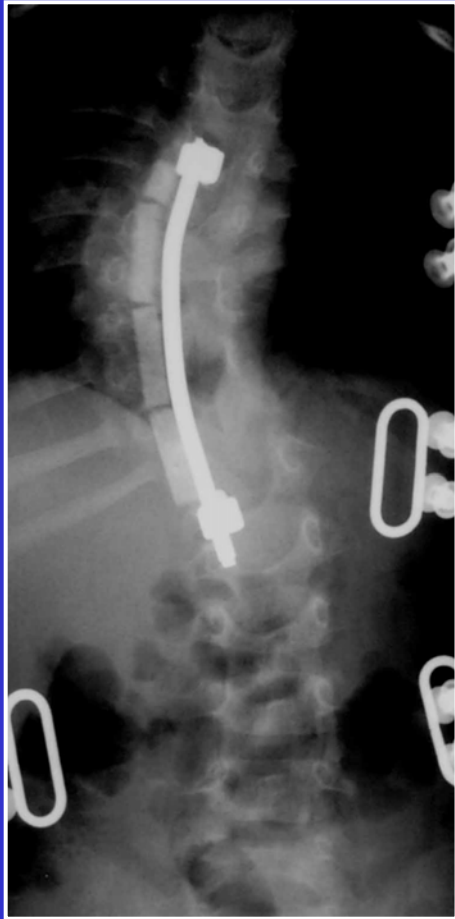
Angle de Cobb s'améliore



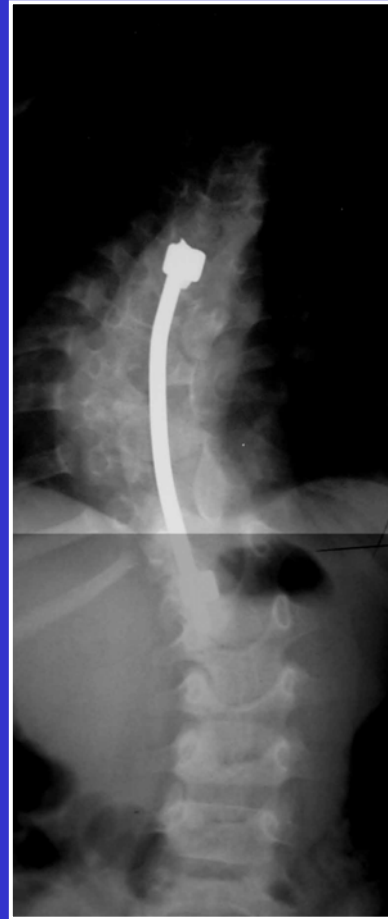




**1 Y 2 M**



**2 Y**



**3 Y**



**9 Y**

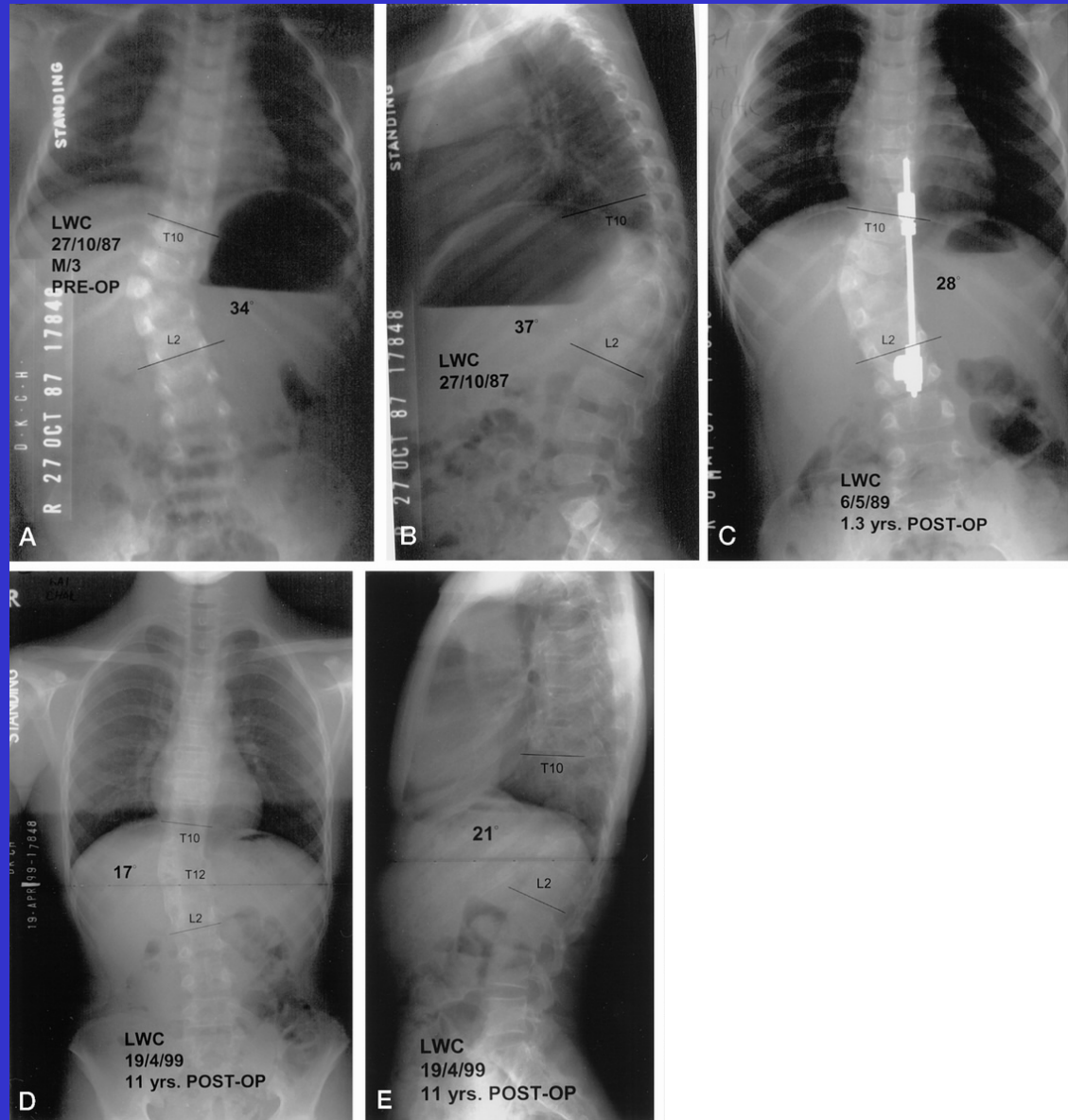




# Ten-Year Follow-up Study of Lower Thoracic Hemivertebrae Treated by Convex Fusion and Concave Distraction

Cheung, Kenneth M. C. FRCS,  
FHKAM(Orth),\*; Zhang, J. G. MD,†;  
Lu, D. S. MD, PhD,\*; K. Luk, Keith D.  
FRCSE, FRCSG, FRACS, MCh(Orth),  
FHKAM(Orth),\* and; Y. Leong, John  
C. OBE, FRCS, FRCSE, FRACS,  
FHKAM(Orth), JP\*

SPINE Volume 27(7) 1 April  
2002 pp 748-753



## OPTIONS THERAPEUTIQUES

Chirurgie « costale »

Epiphysiodèse Convexe

Résection d'hémivertèbre

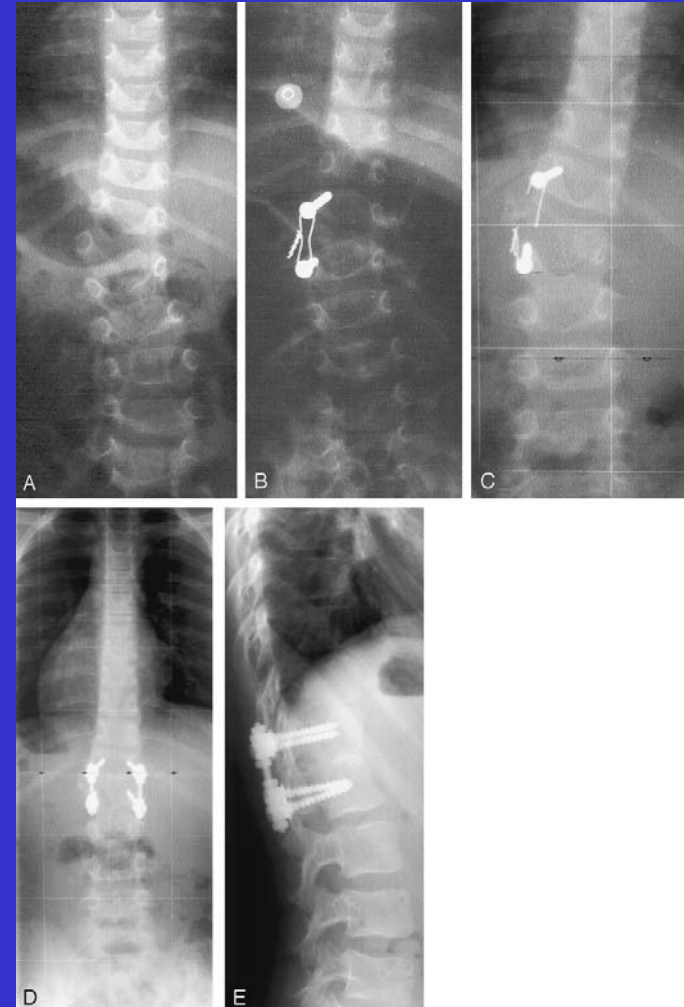


## Posterior Hemivertebra Resection With Transpedicular Instrumentation: Early Correction in Children Aged 1 to 6 Years

Ruf, Michael MD; Harms, Jürgen MD

SPINE Volume 28(18) 15 September 2003 pp 2132-2138

Correction surgery of congenital scoliosis should be performed early, before the development of severe local deformities and secondary structural changes, especially in patients with expected deterioration. Posterior resection of the hemivertebra with transpedicular instrumentation allows for early intervention in very young children. Excellent correction in the frontal and sagittal planes, and a short segment of fusion allows for normal growth in the unaffected parts of the spine.



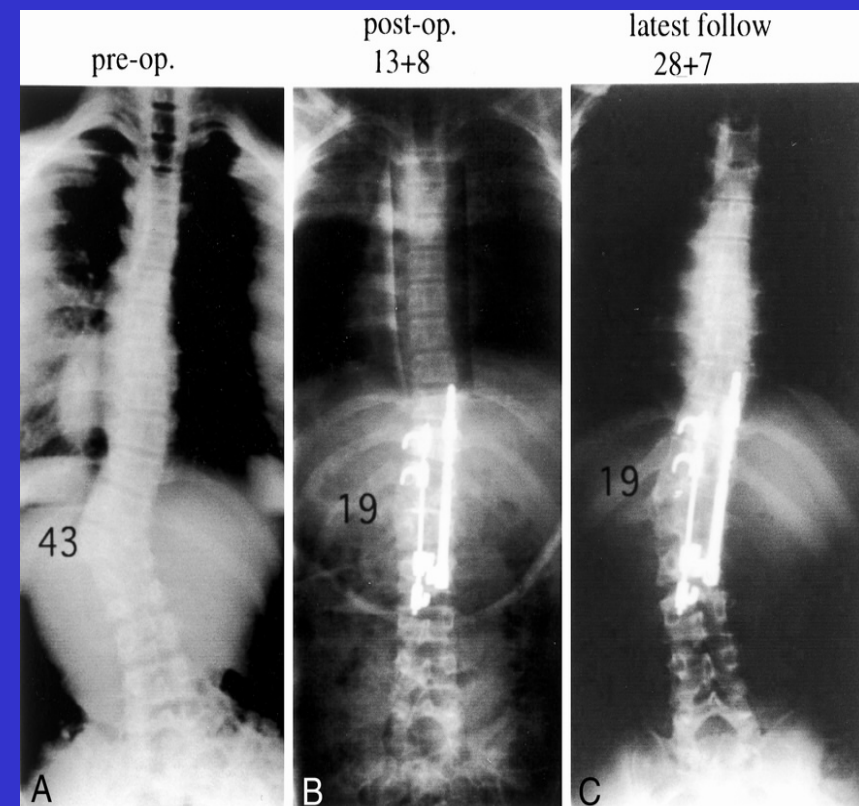
## Single-Stage Excision of Hemivertebrae *Via* the Posterior Approach Alone for Congenital Spine Deformity: Follow-up Period Longer Than Ten Years

Nakamura, Hiroaki MD,\*; Matsuda, Hideki MD,†;  
Konishi, Sadahiko MD,\*; Yamano, Yoshiki

SPINE Volume 27(1) 1 January 2002 pp 110-115

For patients with a thoracolumbar hemivertebra, scoliosis improved from  $49^\circ \pm 6^\circ$  to  $22.3^\circ \pm 3.5^\circ$ , for a 54.3% correction. The correction ratio for kyphosis was 67.4%. Over an average 12.8-year follow up period, loss of scoliotic curvature correction was only  $3.7^\circ$ .

In contrast, the hemivertebral correction ratio for patients with a lumbosacral hemivertebra remained 32.5% because of difficulty using internal fixation associated with patient age. At the most recent follow-up assessment, one patient exhibited deterioration of coronal spinal balance.



# One-Stage Posterior Hemivertebra Resection and Correction Using Segmental Posterior Instrumentation

Shono, Yasuhiro MD, PhD; Abumi, Kuniyoshi MD, PhD;  
Kaneda, Kiyoshi MD, PhD

SPINE Volume 26(7) April 2001 pp 752-757

12 patients

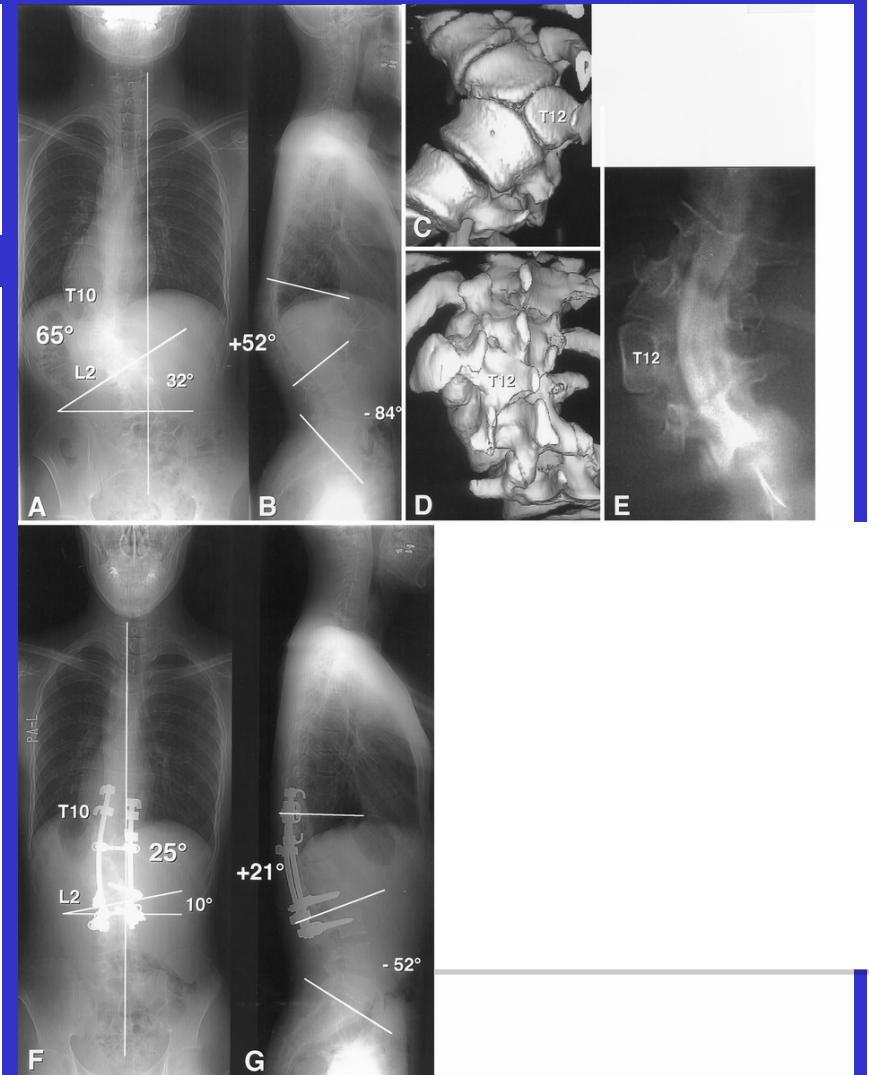
After posterior hemivertebra resection, segmental posterior instrumentation was used for correction of the kyphoscoliotic deformity [CD (4 patients), Kaneda SR (2), and ISOLA (6)].

All 12 patients had kyphoscoliotic deformity.

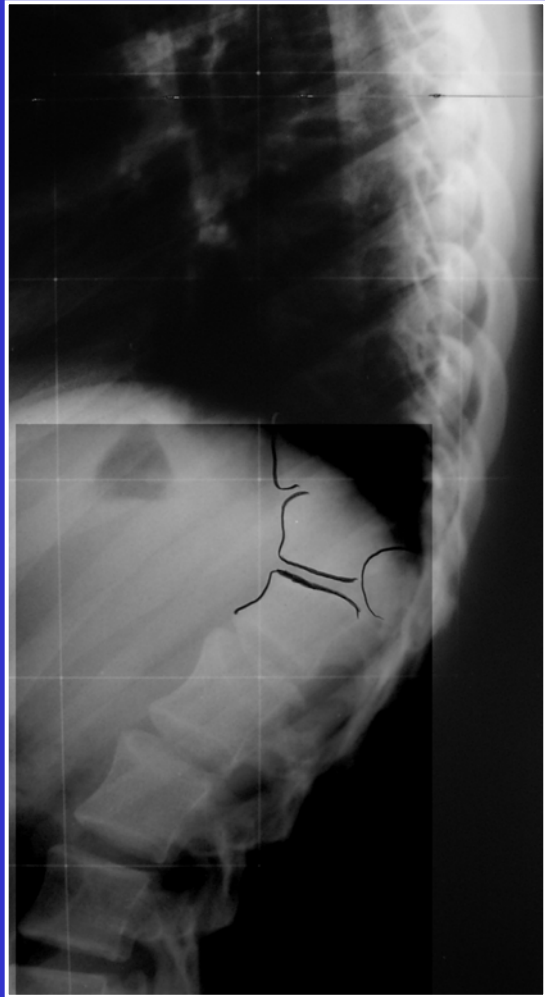
Preoperative scoliosis averaging  $49^\circ$  was corrected to  $18^\circ$  (correction rate, 64%).

Preoperative kyphosis of  $40^\circ$  was corrected to  $17^\circ$  of kyphosis.

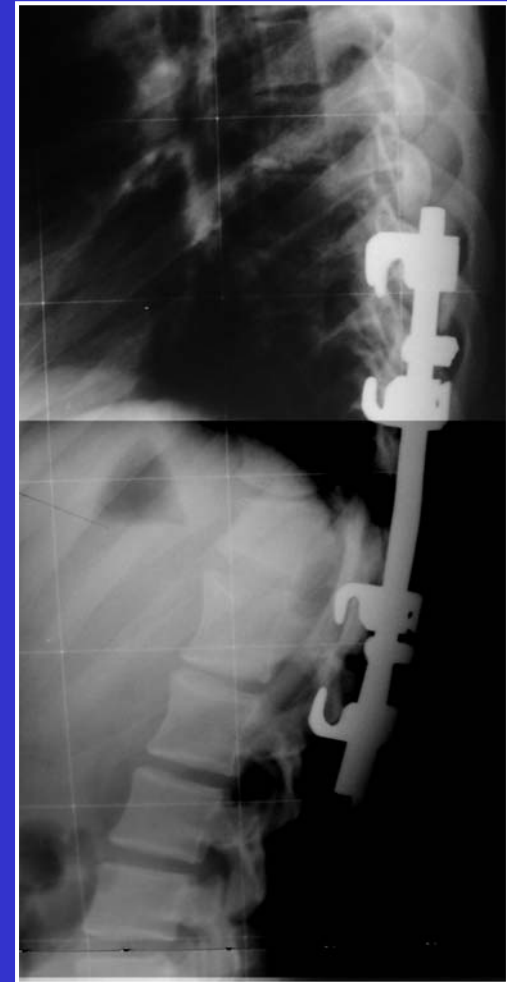
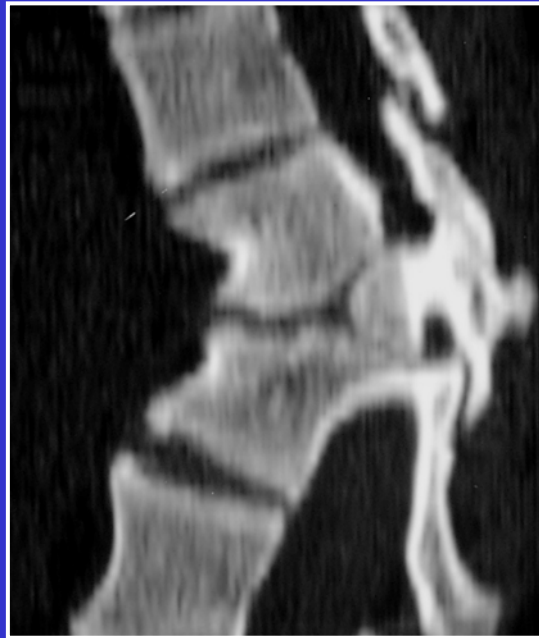
Trunk shift of 23 mm was improved to 3 mm. Correction loss was  $2^\circ$  in the frontal plane and  $3^\circ$  in the sagittal plane, and no patients showed more than  $5^\circ$  of correction loss.







12 Y



15 Y



## Transpedicular Eggshell Osteotomies for Congenital Scoliosis Using Frameless Stereotactic Guidance

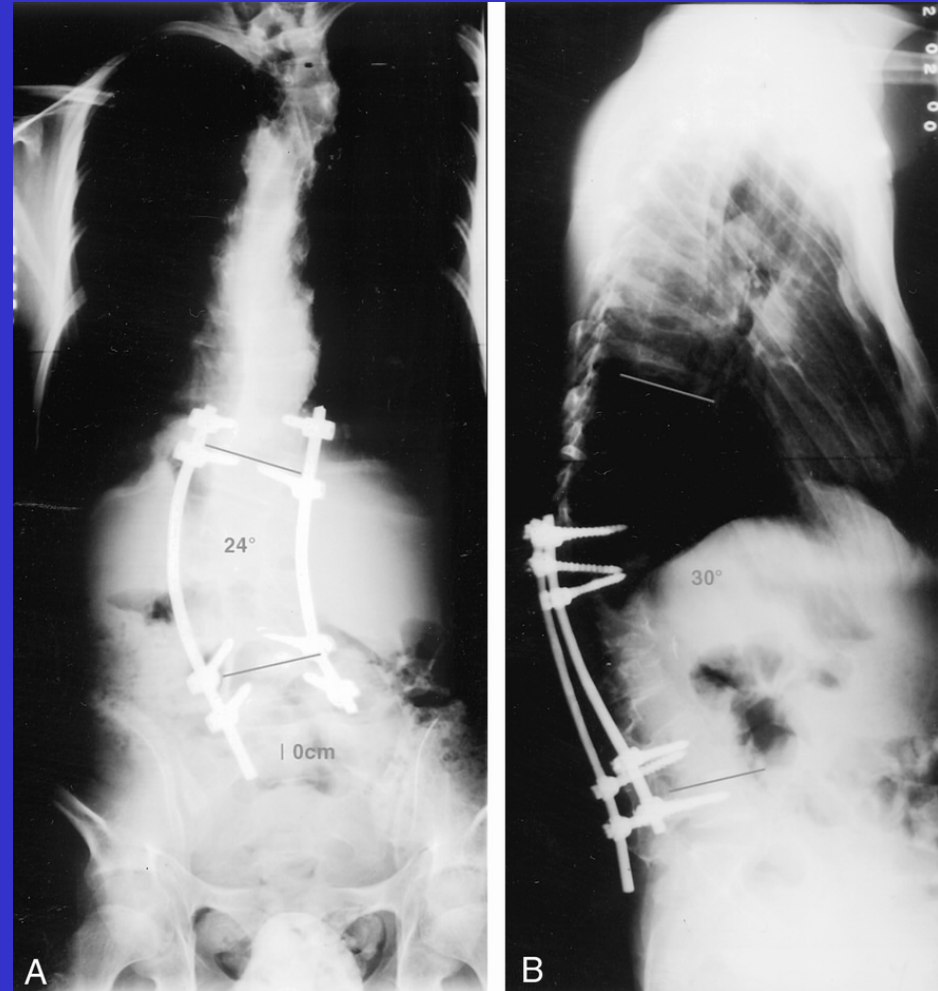
Mikles, Mark R. MD; Graziano, Gregory P. MD;  
Hensinger, and Robert N. MD

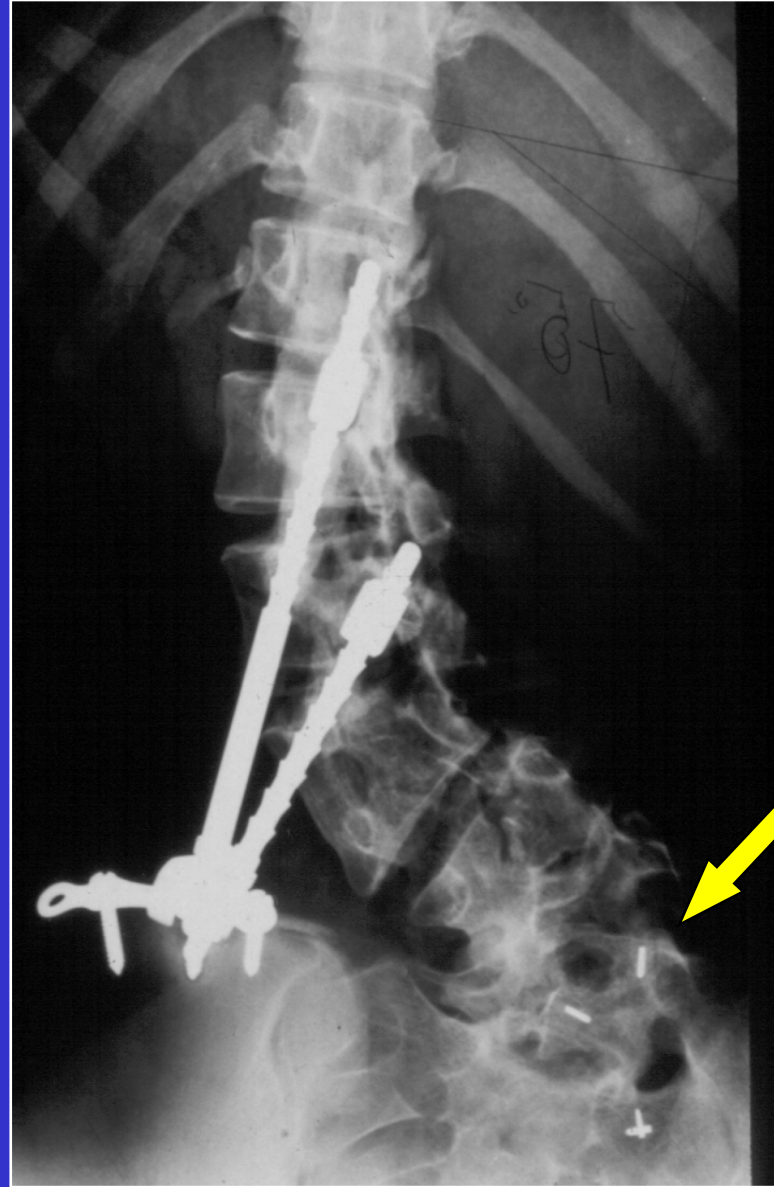
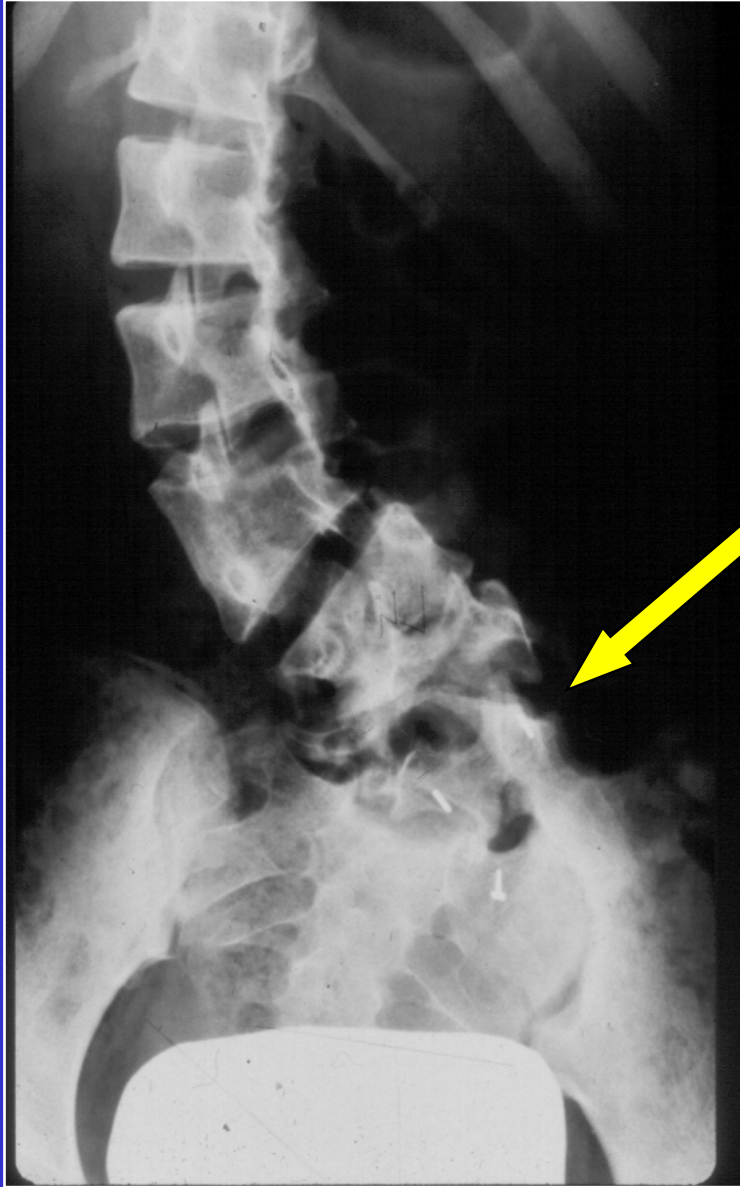
SPINE Volume 26(20) 15 October 2001 pp  
2289-2296

Three older patients who had congenital scoliosis.....

The average coronal correction of the major curve was  $28.7^\circ$  (range,  $22-33^\circ$ ). The average correction of the plumb line or lateral displacement from the center of the trunk was 4.8 cm (range, 3–7.5 cm).

A significant kyphotic deformity was corrected  $38^\circ$ , and a pelvic tilt was reduced from 7 to 3 cm.







## **Hemivertebral Excision for Congenital Scoliosis in Very Young Children**

[Klemme, William](#) R. M.D.; Polly, David W. Jr. M.D.; Orchowski, Joseph R. M.D.

SPINE Volume 21(6) November/December 2001 pp 761-764

## **Excision of hemivertebrae in the management of congenital scoliosis involving the thoracic and thoracolumbar spine**

[Deviren, V.](#); Berven, S.; Smith, J. A.; Emami, A.; Hu, S. S.; Bradford, D. S.

*Journal of Bone and Joint Surgery* Volume 83-B(4) May 2001 pp 496-500

## **Simultaneous Anterior and Posterior Hemivertebra Excision**

[Section I: Symposium: Spinal Deformities and Pediatric Orthopaedics: A Tribute to John E. Hall:  
The Classic]

[Lazar, Richard D.](#) MD\*; Hall, John E. MD\*\*

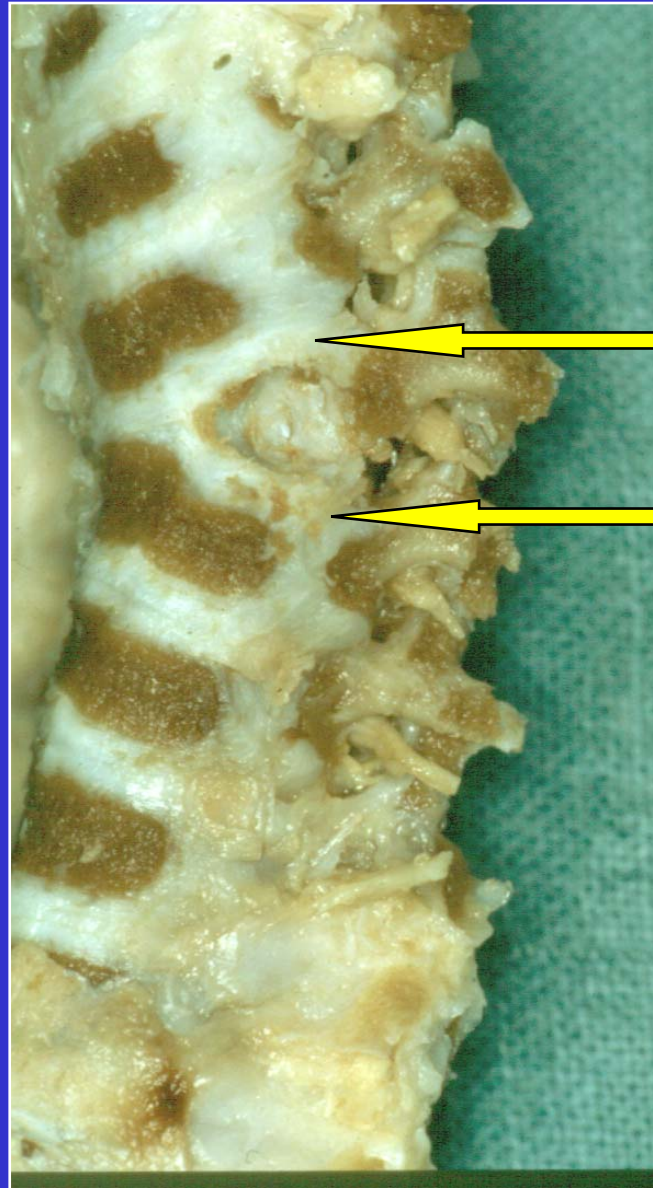
SPINE Volume 1(364) July 1999 pp 76-84

## **Excision of Hemivertebrae and Wedge Resection in the Treatment of Congenital Scoliosis**

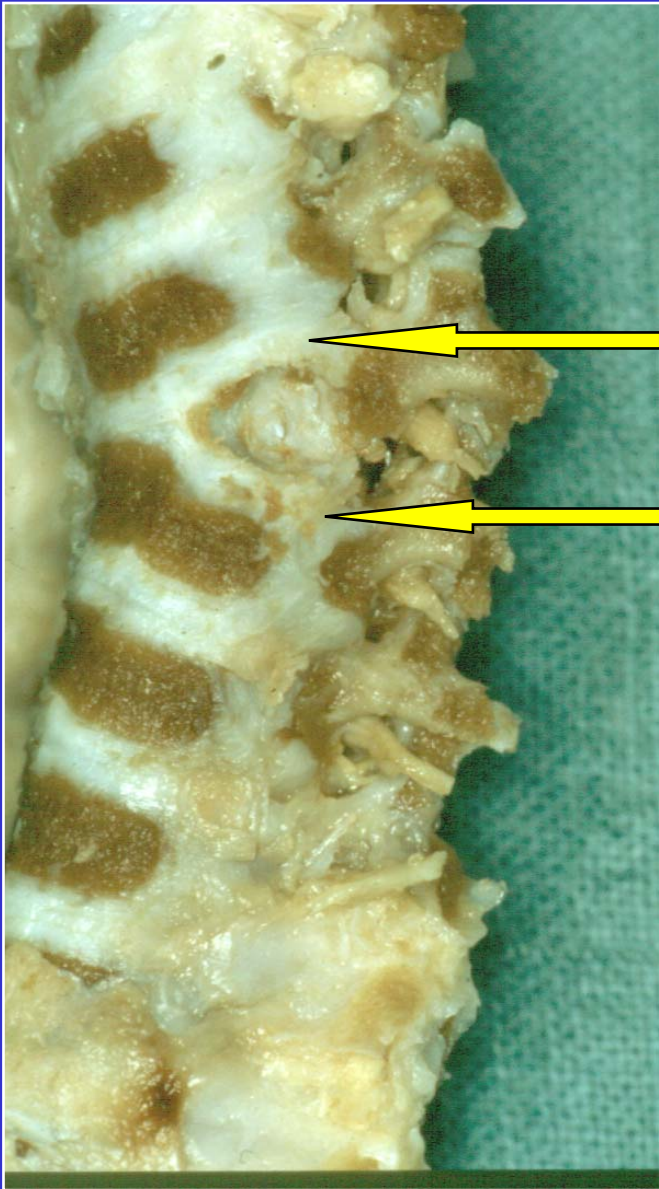
[Holte, David C.](#); Winter, Robert B.; Lonstein, John E.; Denis, Francis

*The Journal of Bone and Joint Surgery* Volume 77-A(2) February 1995 pp 159-171

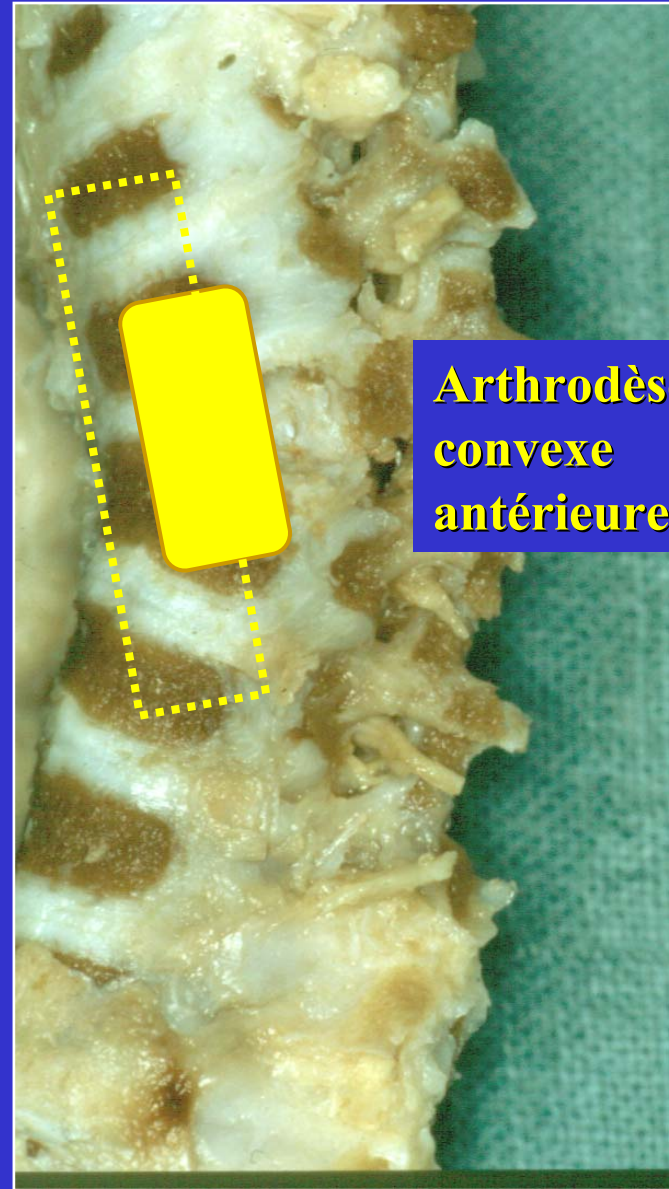




**Structures de  
croissance**



**Structures de croissance**



**Arthrodèse convexe antérieure**

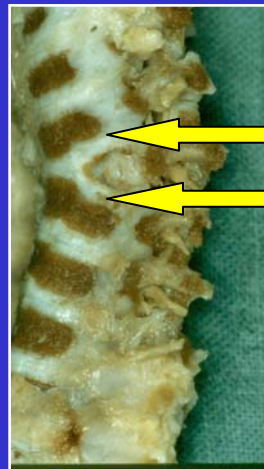


Sur une période de vingt ans 79 des 220 hémivertèbres suivies par nos soins ont bénéficié d'une résection

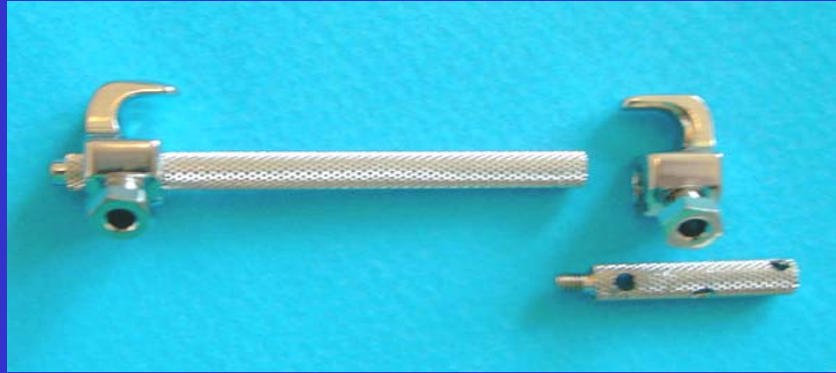
## 79 RESECTIONS d' HEMIVERTEBRES

### Technique

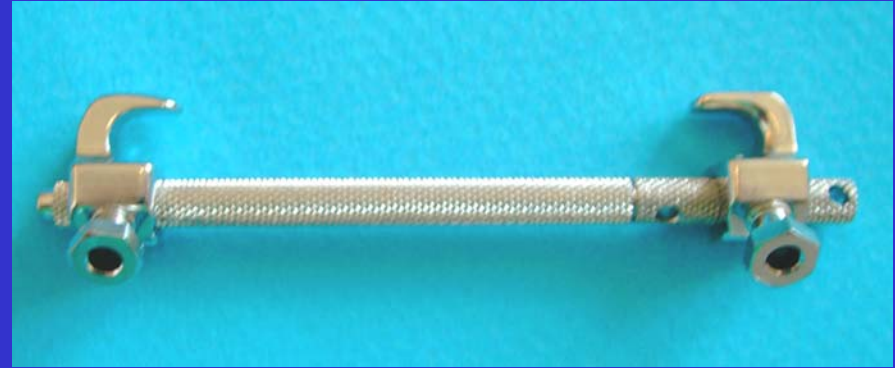
Abord antérieur et postérieur en un temps opératoire avec arthrodèse convexe antérieure et postérieure et instrumentation convexe postérieure en un seul temps utilisant le Baby CD + corset plâtré pendant 6 mois



Structures de croissance à réséquer



1



2

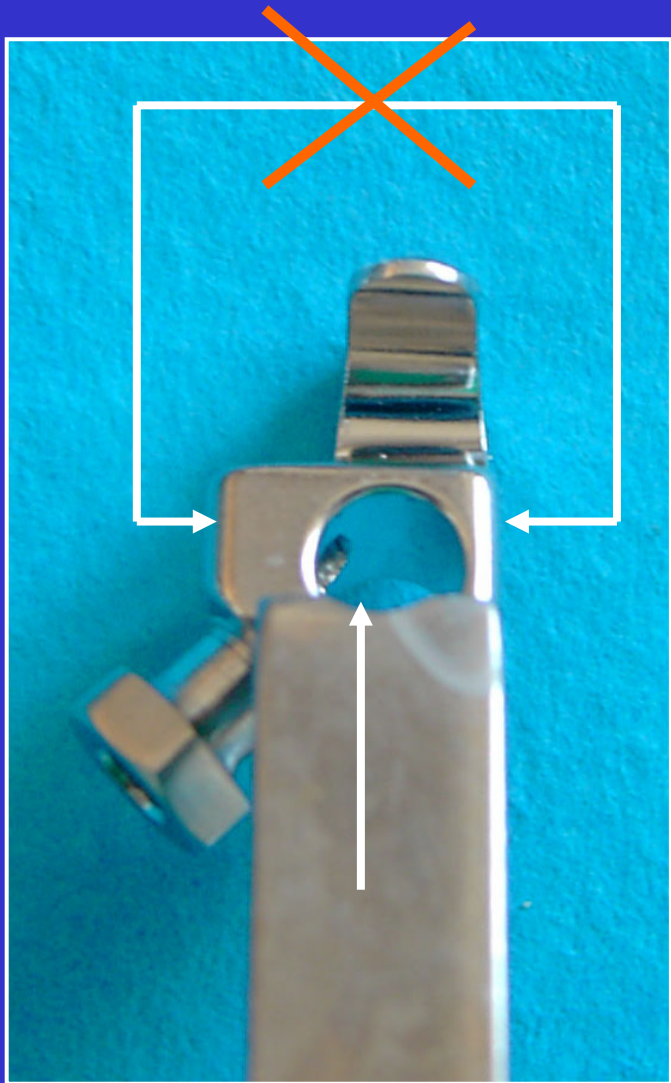


3



4

BABY CD



# SCOLIOSES

Courbure segmentaire (°)

Courbure principale globale (°)

Contre courbure craniale (°)

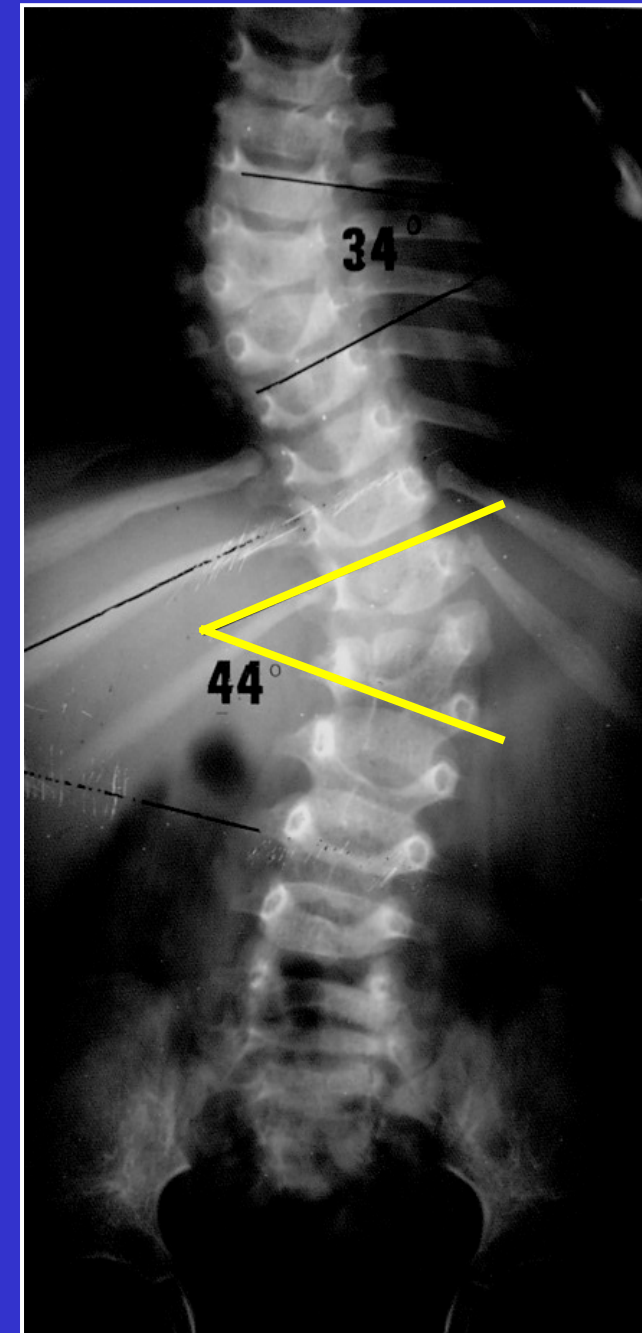
Contre courbure caudale (°)

Translation gravitaire (mm)

Translation gravitaire (%)

Translation vraie (mm)

Translation vraie (%)





# SCOLIOSES

Courbure segmentaire (°)

**Courbure principale globale (°)**

Contre courbure craniale (°)

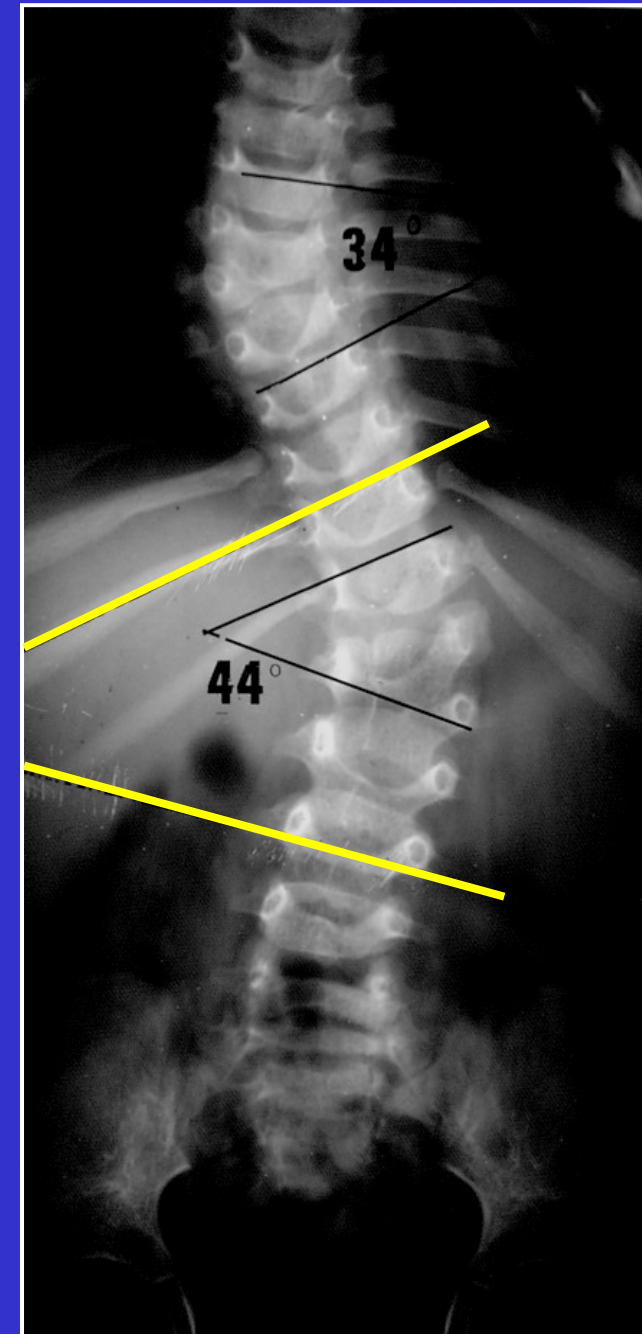
Contre courbure caudale (°)

Translation gravitaire (mm)

Translation gravitaire (%)

Translation vraie (mm)

Translation vraie (%)



# SCOLIOSES

Courbure segmentaire (°)

Courbure principale globale (°)

**Contre courbure craniale (°)**

Contre courbure caudale (°)

Translation gravitaire (mm)

Translation gravitaire (%)

Translation vraie (mm)

Translation vraie (%)





# SCOLIOSES

Courbure segmentaire (°)

Courbure principale globale (°)

Contre courbure craniale (°)

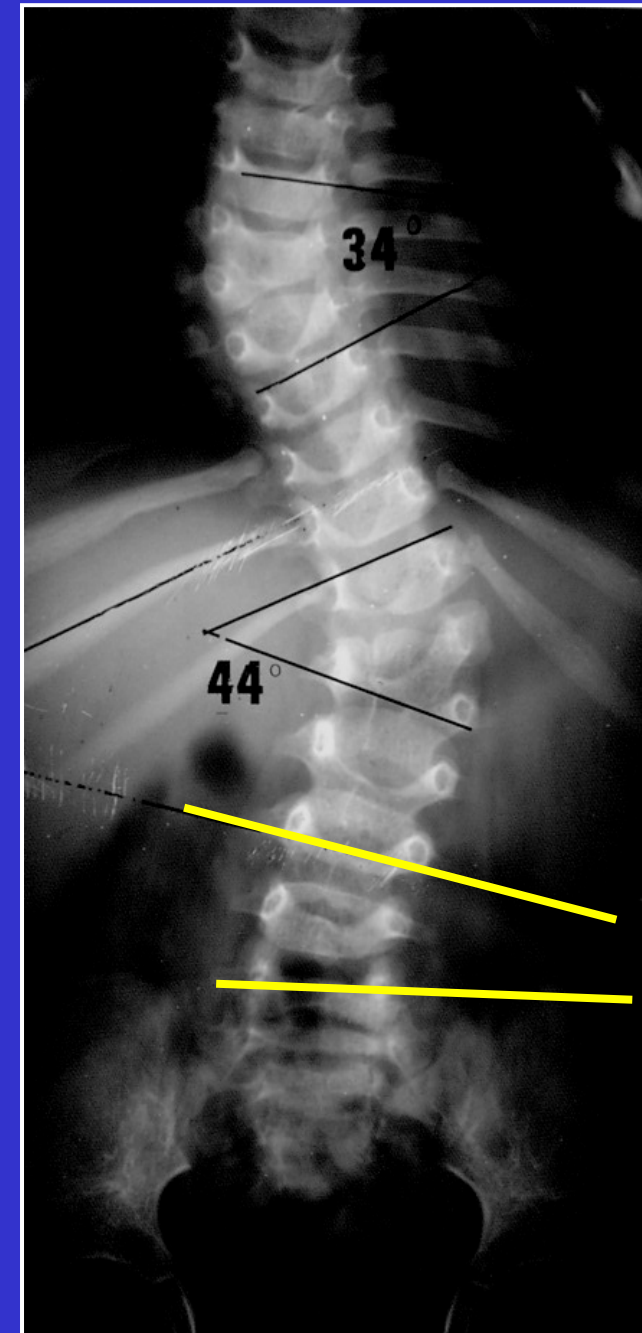
**Contre courbure caudale (°)**

Translation gravitaire (mm)

Translation gravitaire (%)

Translation vraie (mm)

Translation vraie (%)



# TRANSLATION DU TRONC

Courbure segmentaire (°)

Courbure principale globale (°)

Contre courbure craniale (°)

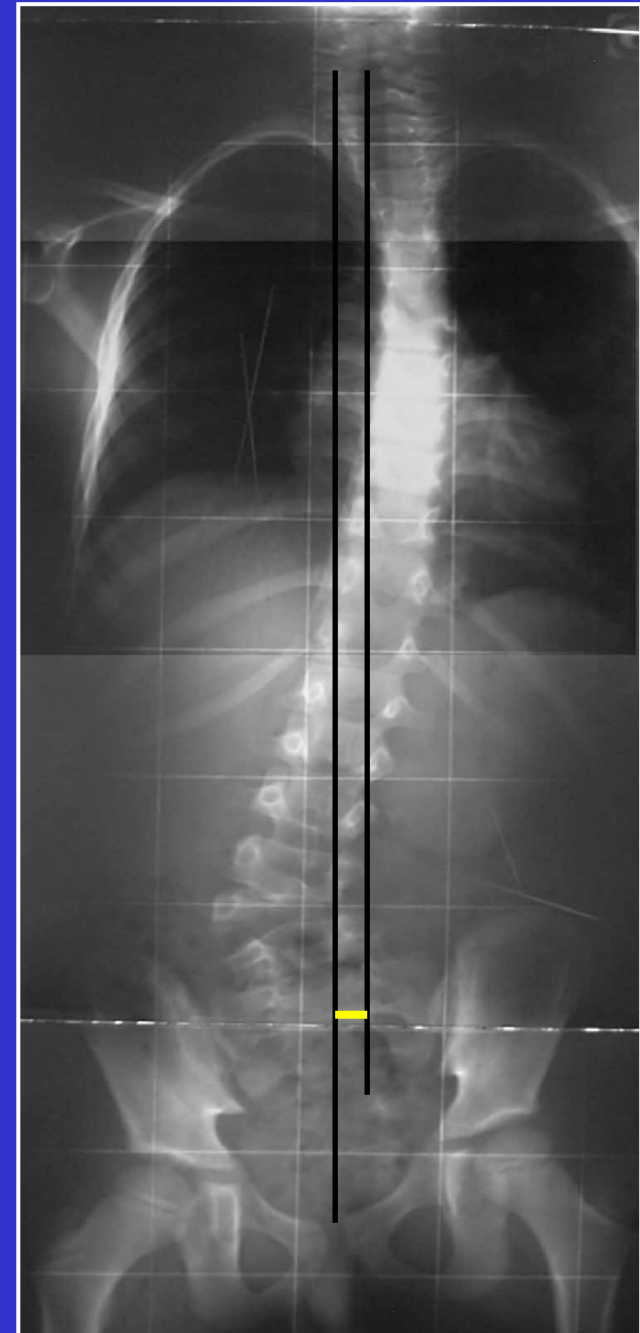
Contre courbure caudale (°)

**Translation gravitaire (mm)**

**Translation gravitaire (%)**

Translation vraie (mm)

Translation vraie (%)



# TRANSLATION DU TRONC

Courbure segmentaire (°)

Courbure principale globale (°)

Contre courbure craniale (°)

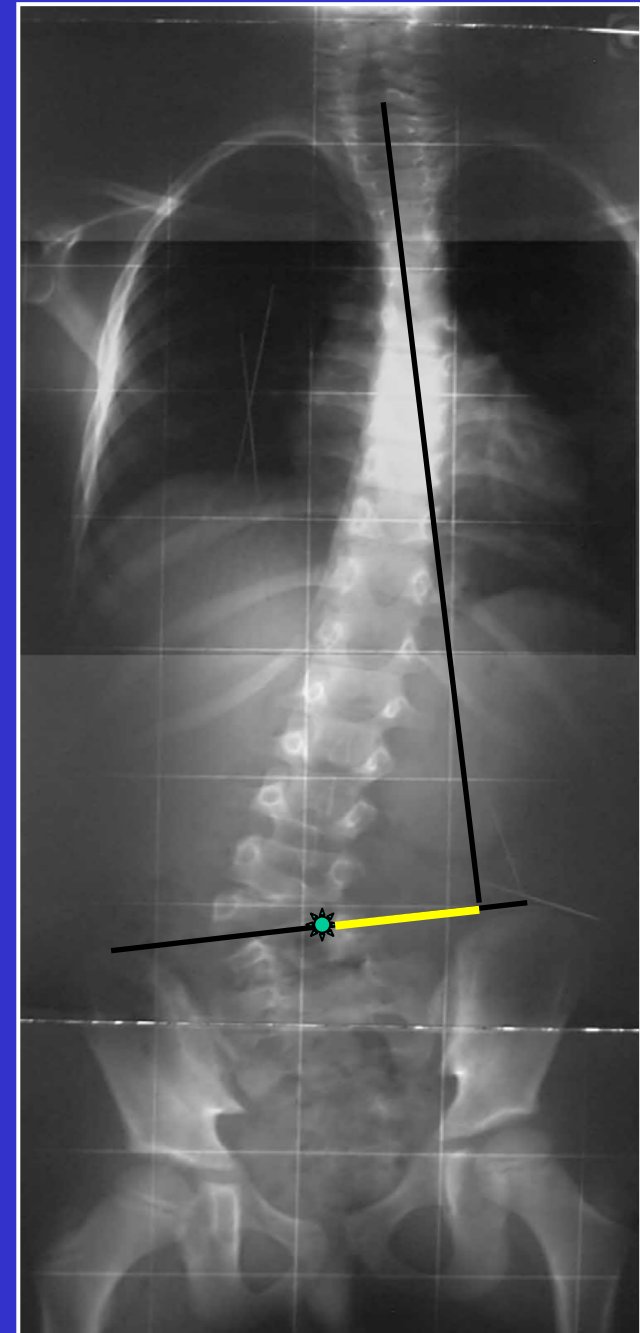
Contre courbure caudale (°)

Translation gravitaire (mm)

Translation gravitaire (%)

**Translation vraie (mm)**

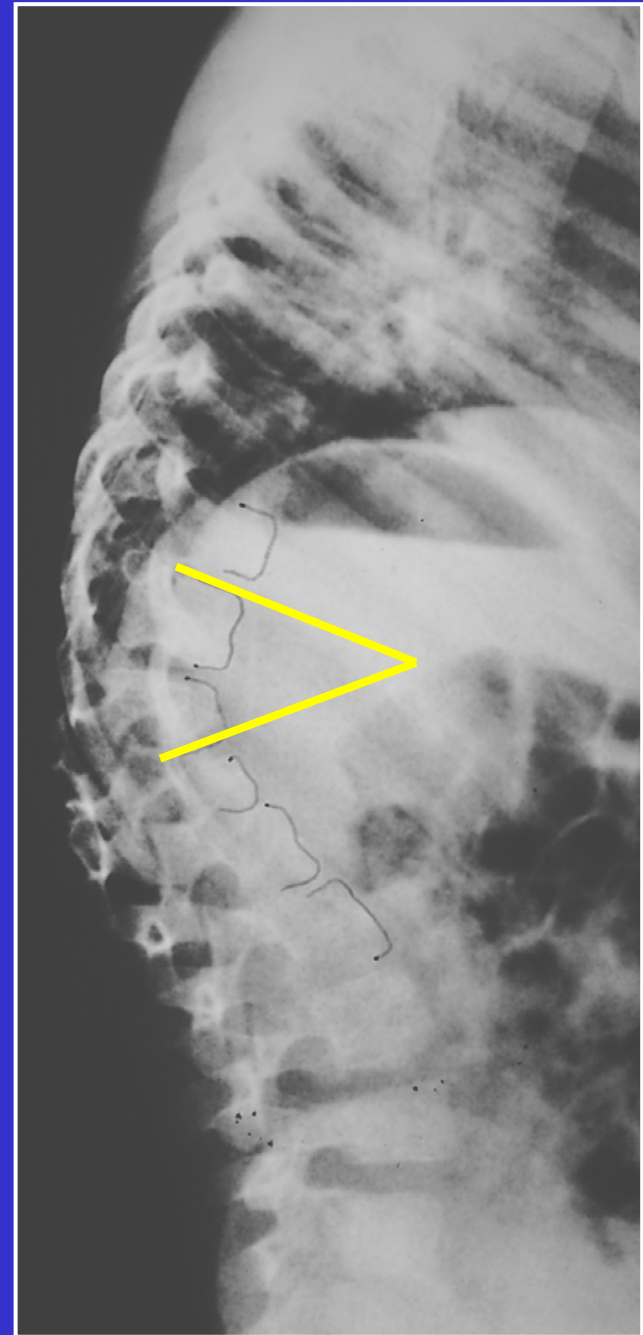
**Translation vraie (%)**



# CYPHOSE

Courbure segmentaire (°)

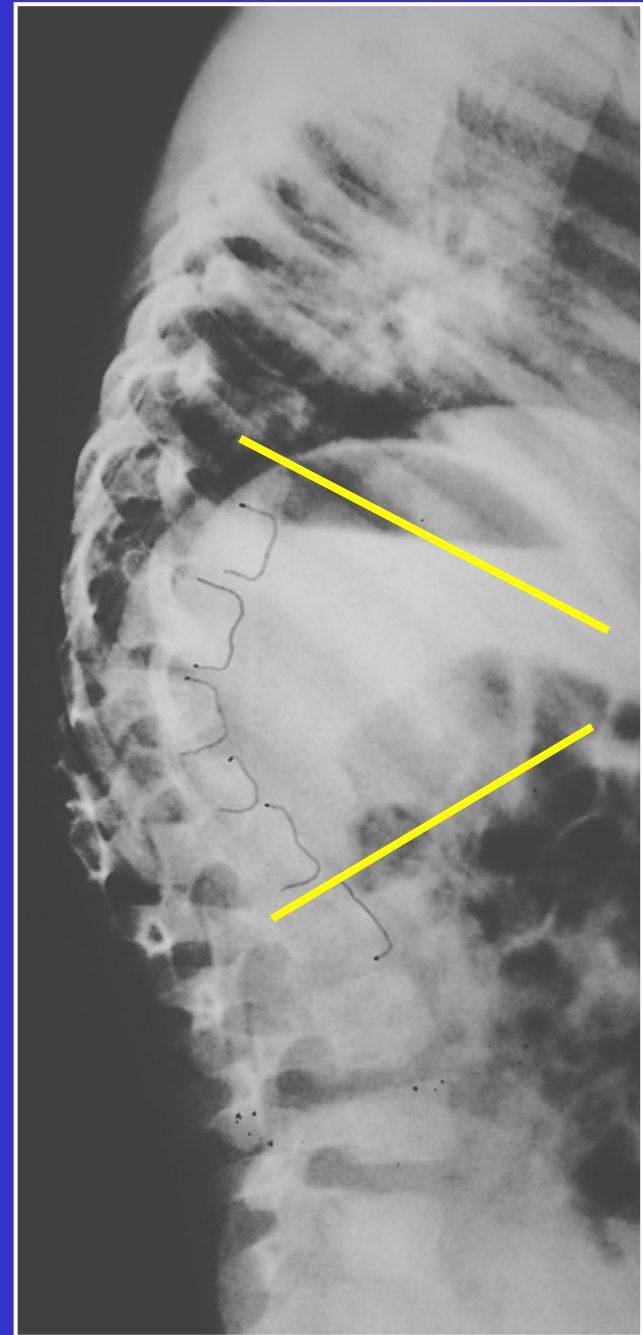
Courbure principale globale (°)



# CYPHOSE

Courbure segmentaire (°)

Courbure principale globale (°)



## 36 HEMIVERTEBRES THORACO-LOMBAIRES

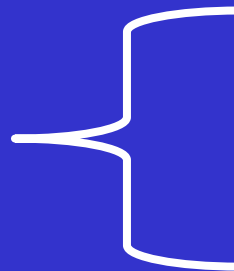
LOCALISATION ENTRE T9 ET L2

22 DROITES 14 GAUCHES

SEXE

19 FILLES 17 GARCONS

SEGMENTATION



12 LIBRES

23 SEMI SEGMENTEES

1 NON SEGMENTEE

AGE A L'OPERATION

3.5 Y (12M - 9.8Y)

SUIVI

5A 10M (10M - 14A)

DUREE DE L'INTERVENTION

282' (210' - 345')

PERTE D'HEMOGLOBINE

3.7 g/dl (1.4 - 7)

4 Patients transfusés



<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9mm(1-30)	10.8mm (1-43)	8.7mm (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5mm (1-43)	13.6mm (0-47)	10.5mm(0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)

<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9 <sub>mm</sub> (1-30)	10.8 <sub>mm</sub> (1-43)	8.7 <sub>mm</sub> (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5 <sub>mm</sub> (1-43)	13.6 <sub>mm</sub> (0-47)	10.5 <sub>mm</sub> (0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)

<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9 <sub>mm</sub> (1-30)	10.8 <sub>mm</sub> (1-43)	8.7 <sub>mm</sub> (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5 <sub>mm</sub> (1-43)	13.6 <sub>mm</sub> (0-47)	10.5 <sub>mm</sub> (0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)

<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9 <sub>mm</sub> (1-30)	10.8 <sub>mm</sub> (1-43)	8.7 <sub>mm</sub> (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5 <sub>mm</sub> (1-43)	13.6 <sub>mm</sub> (0-47)	10.5 <sub>mm</sub> (0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)

<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9 <sub>mm</sub> (1-30)	10.8 <sub>mm</sub> (1-43)	8.7 <sub>mm</sub> (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5 <sub>mm</sub> (1-43)	13.6 <sub>mm</sub> (0-47)	10.5 <sub>mm</sub> (0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)



<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9 <sub>mm</sub> (1-30)	10.8 <sub>mm</sub> (1-43)	8.7 <sub>mm</sub> (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5 <sub>mm</sub> (1-43)	13.6 <sub>mm</sub> (0-47)	10.5 <sub>mm</sub> (0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)

<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	34.4°(16-62)	16.2° (1-44)	11.9° (0-42)
Courbure principale globale (°)	40.6° (24-66)	22.8° (2-46)	25.4° (0-76)
Contre courbure craniale (°)	13.7° (0-34)	10.2° (0-36)	10.4° (0-34)
Contre courbure caudale (°)	19.8° (1-48)	9.9° (0-27)	10.2° (0-43)
Translation gravitaire (mm)	10.9 <sub>mm</sub> (1-30)	10.8 <sub>mm</sub> (1-43)	8.7 <sub>mm</sub> (0-20)
Translation gravitaire (%)	11.1%(0.9-33.7)	10.2% (0.9-32)	7.2% (0-17.2)
Translation vraie (mm)	13.5 <sub>mm</sub> (1-43)	13.6 <sub>mm</sub> (0-47)	10.5 <sub>mm</sub> (0-30)
Translation vraie (%)	14% (1.1-47.8)	13% (0-34.8)	8.3%(0-21.9)

<b>CYPHOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	20.5°(-11/78)	9.3°(-5/27)	14.4° (-10/40)
Courbure principale globale (°)	34.3°(2/70)	30.5° (6/53)	35.9° (10/66)

<b>CYPHOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	20.5°(-11/78)	9.3°(-5/27)	14.4° (-10/40)
Courbure principale globale (°)	34.3°(2/70)	30.5° (6/53)	35.9° (10/66)

## **Malformations congénitales associées**

**3 agénésies rénales et 1 rein en fer à cheval**

**VATER (défaut du septum ventriculaire, atrésie de l'oesophage avec fistule oeso-trachéale, anomalies des arcs branchiaux**

**VATER (ventricule droit unique opéré, défaut du septum atrial, imperforation anale avec fistule vaginale, dilatation pyélocalicielle bilatérale)**

**dextrocardie, imperforation anale**

**cryptorchidie**

**angiome lombaire, agénésie cutanée dorsale congénitale**

**hypospadias**

**retard intellectuel**

**imperforation anale**

**stenose hypertrophique du pylore**

**défaut de septum atrial et ventriculaire, stenose de l'artère pulmonaire**

**transposition des gros vaisseaux**

**reflux vésico-urétéral**

**2 moelles fixées, 1 syringomyélie**

## **CHIRURGIE ADDITIONNELLE**

**Révision de la greffe pour pseudarthrose et arthrodèse définitive**

**10 ablations de matériel**

**Distraction par tige sous cutanée sans arthrodèse pour aggravation angulaire**

**Arthrodèse postérieure pour rupture d'une tige de Harrington**

**Révision de greffe pour pseudarthrose (addition d'une 2ème tige), ablation de la tige convexe pour sepsis, ablation de la tige concave et arthrodèse définitive (CD)**

**Tige sous cutanée en distraction sans arthrodèse suivi de 3 allongements**

**Tige sous cutanée en distraction sans arthrodèse, allongement de tige, arthrodèse postérieure (CD), ablation de matériel**

**Révision de greffe pour pseudarthrose**

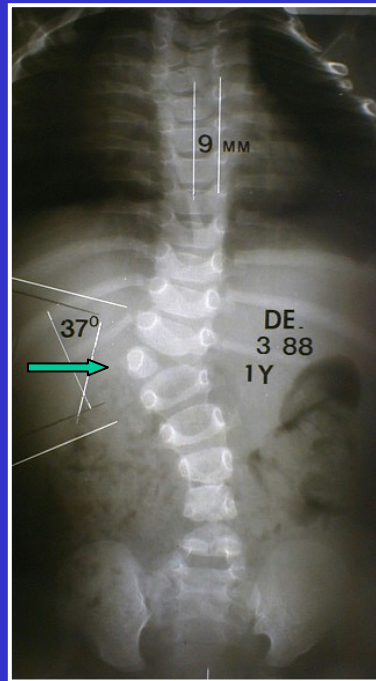
**Exérèse d'une bande fibreuse cicatricielle sur la prise de greffe de péroné**

**Hernie de paroi abdominale, ablation de matériel, ostéotomie vertébrale, arthrodèse postérieure définitive (CD) pour cyphose**

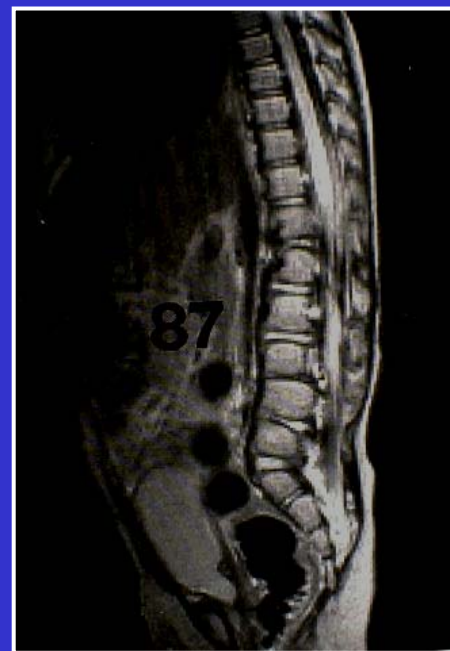
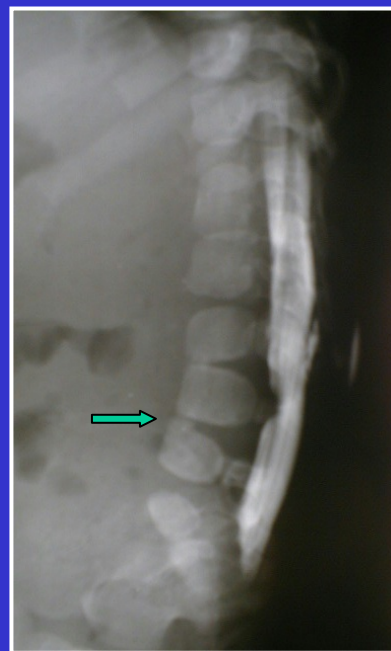
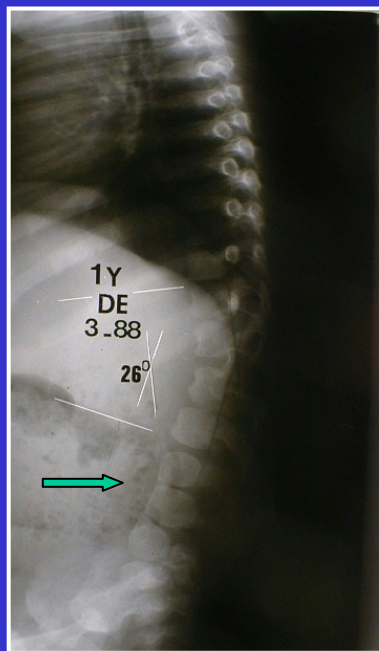
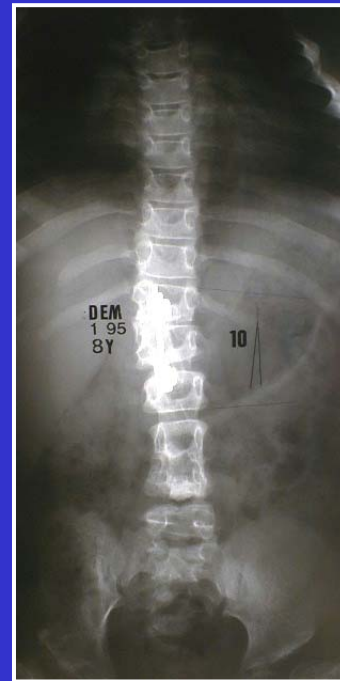
**Changement de crochet**

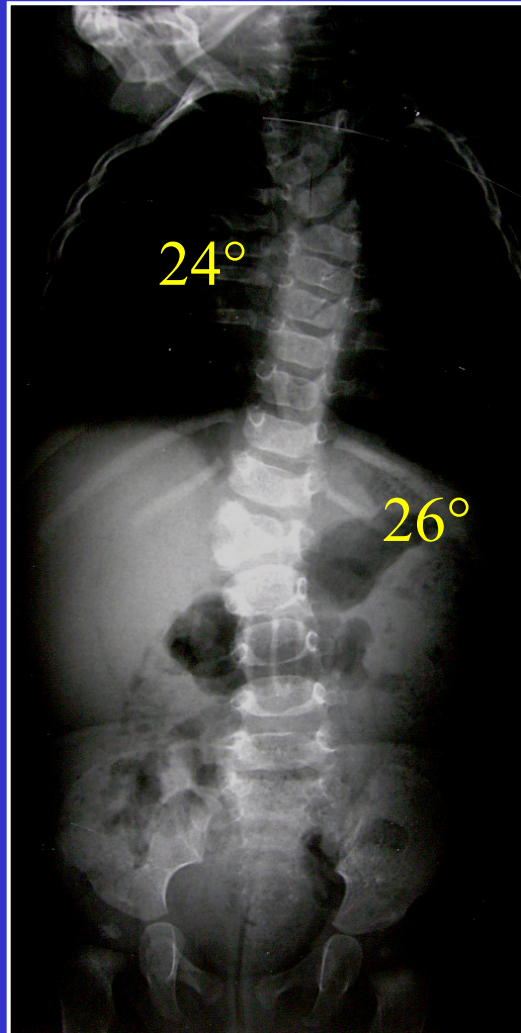


1 A

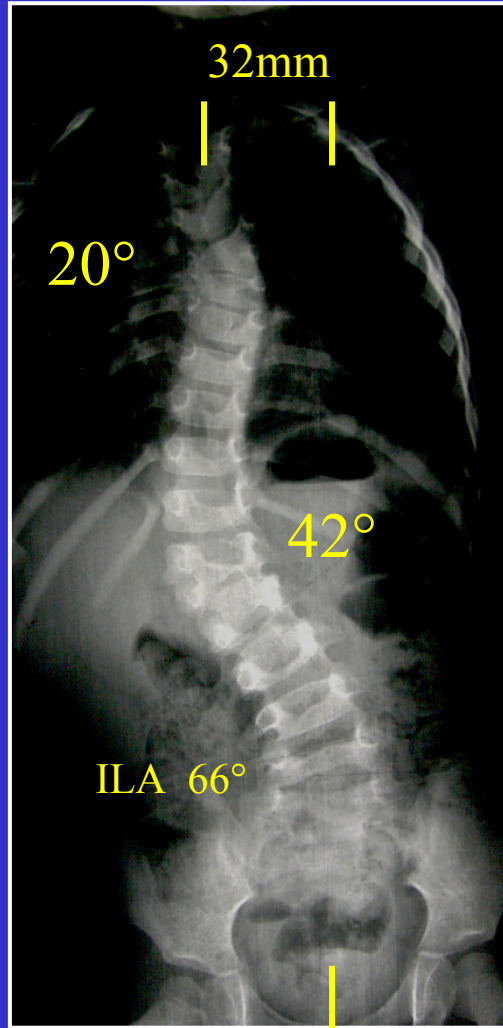


8 A

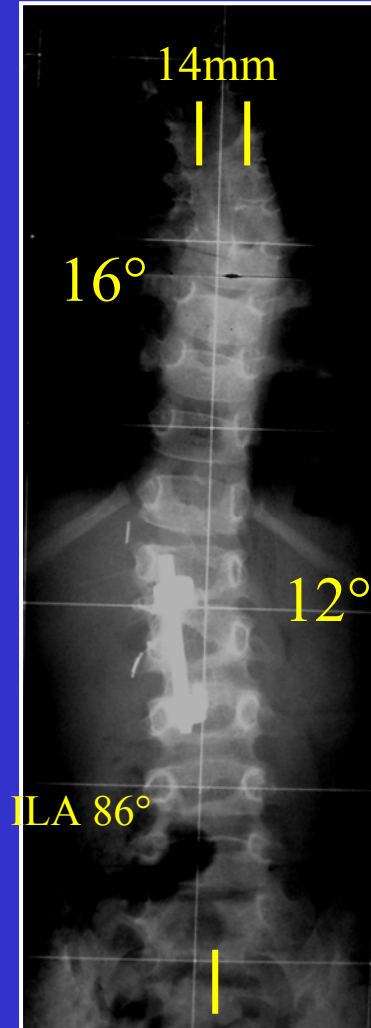




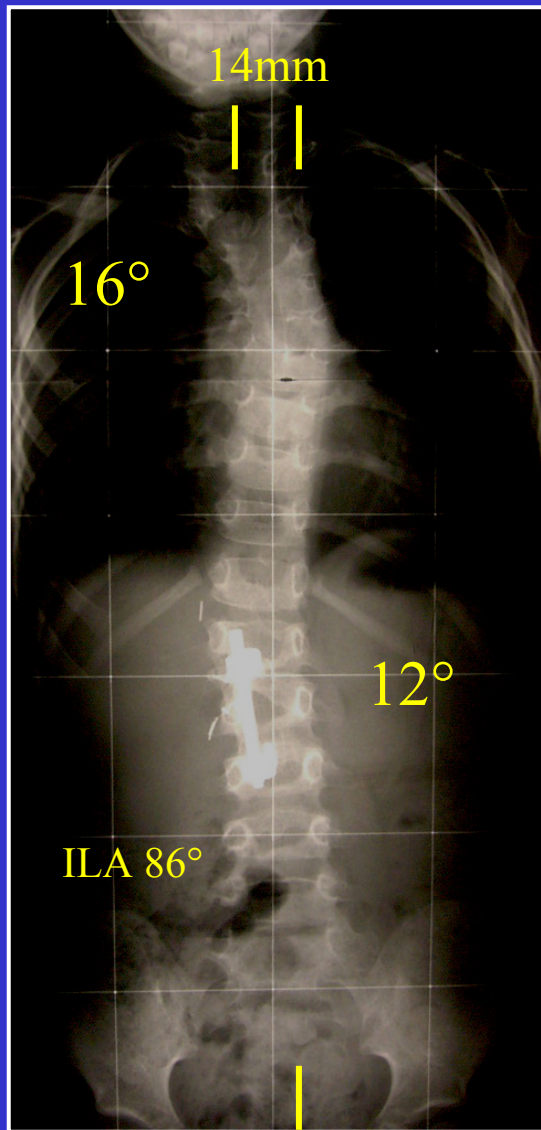
7 Mois



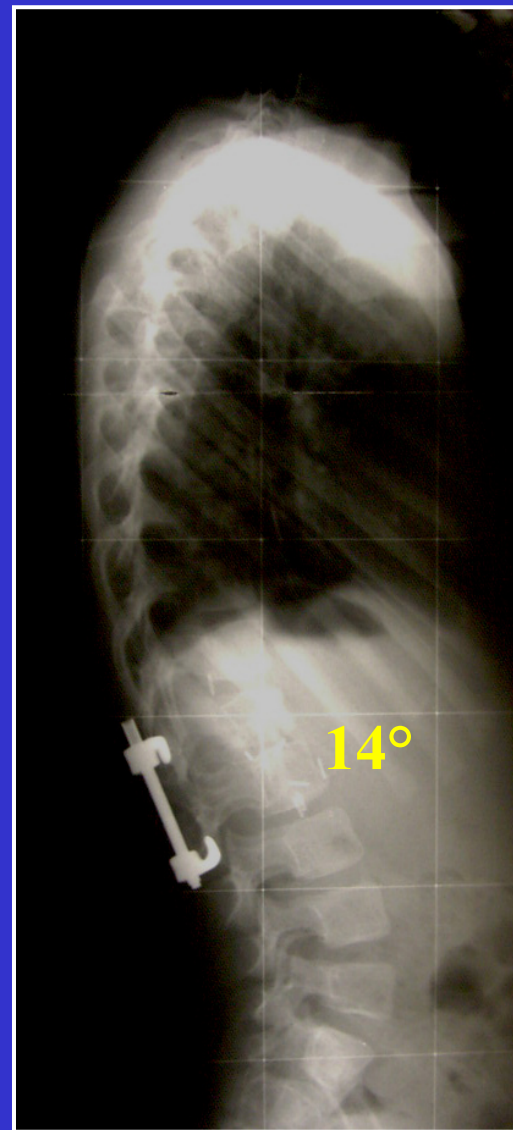
18 Mois



4 A 6 M

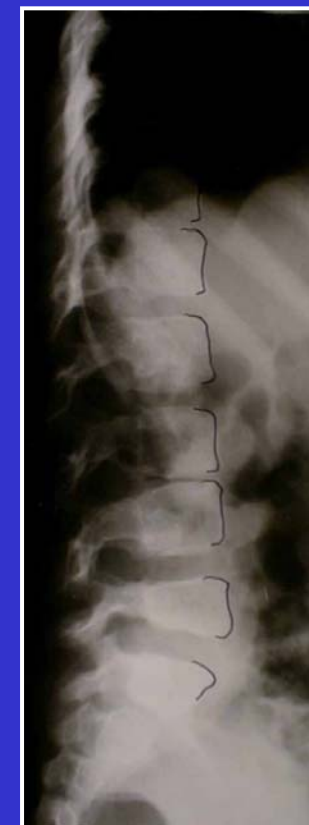
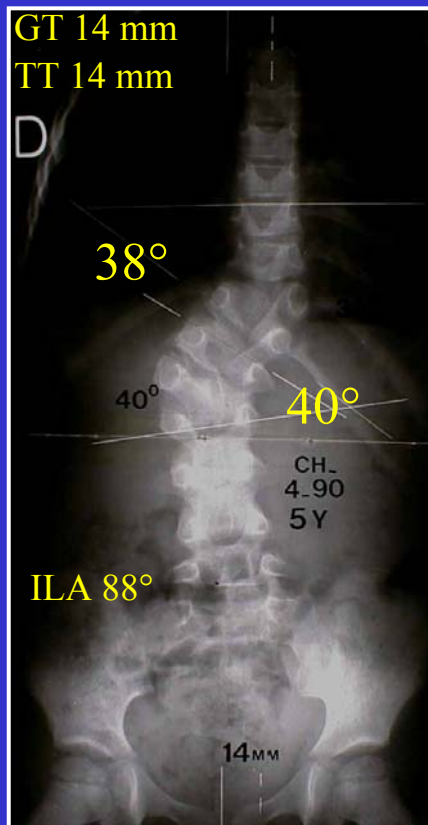


4 A 6 M



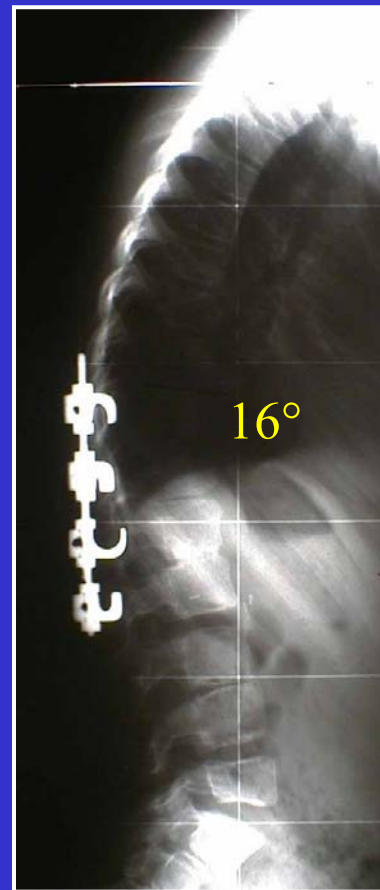
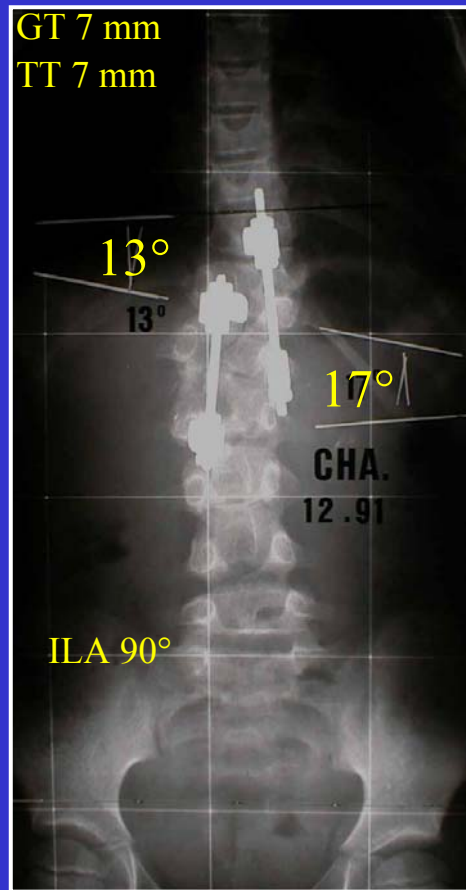
4 A 6 M

# H.V. Libre T11 T12 gauche + H.V. Libre L1 L2 droite

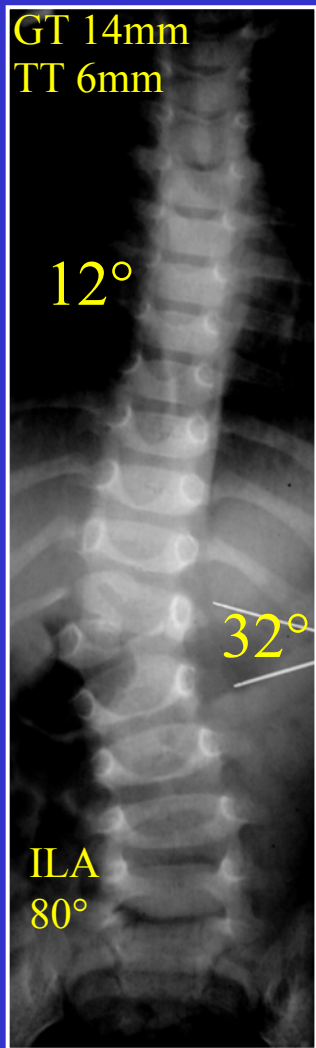


5 A

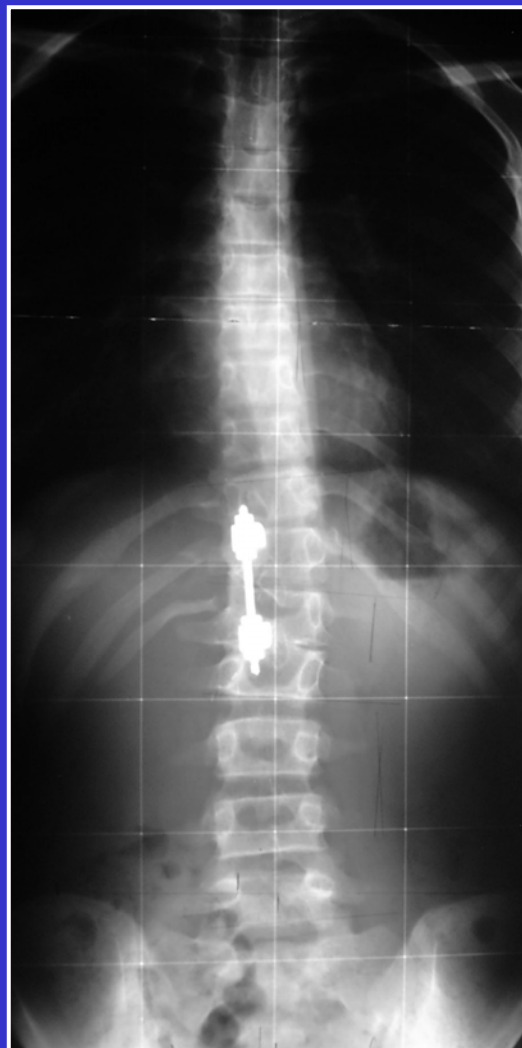




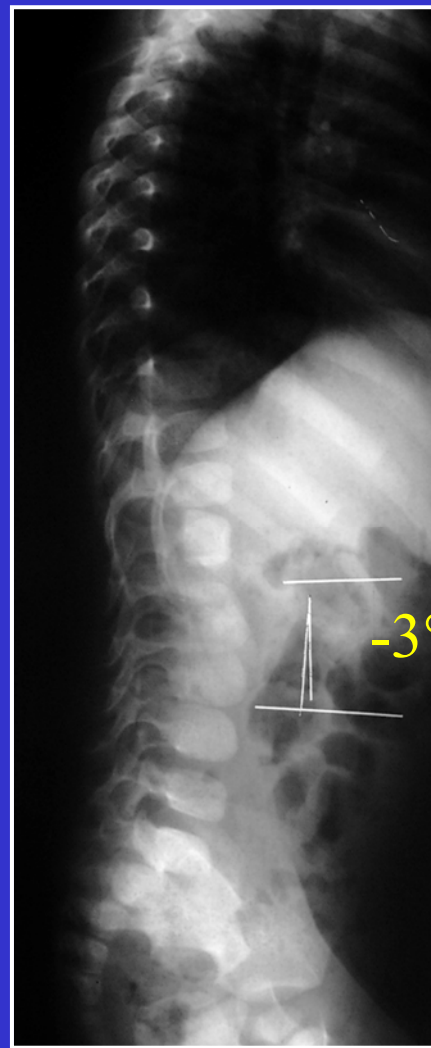
7 A



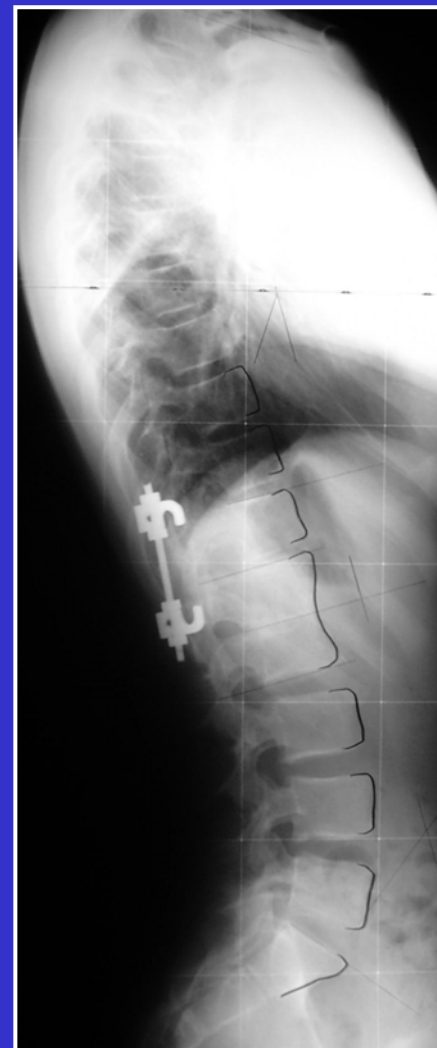
1A 10M



13A 10M



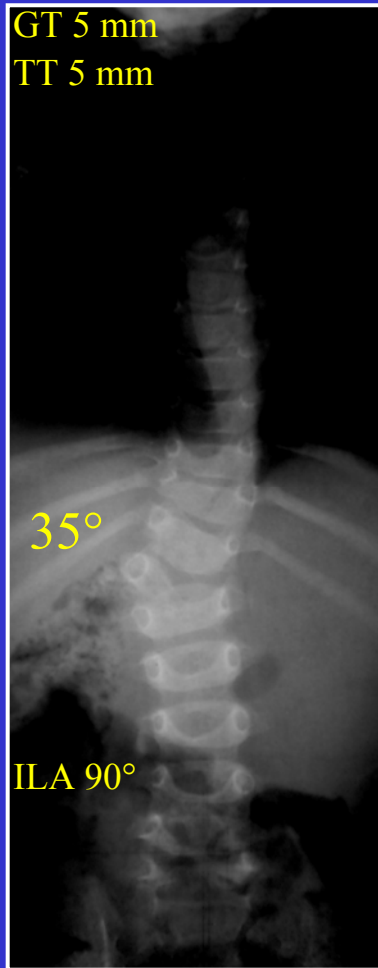
1A 10M



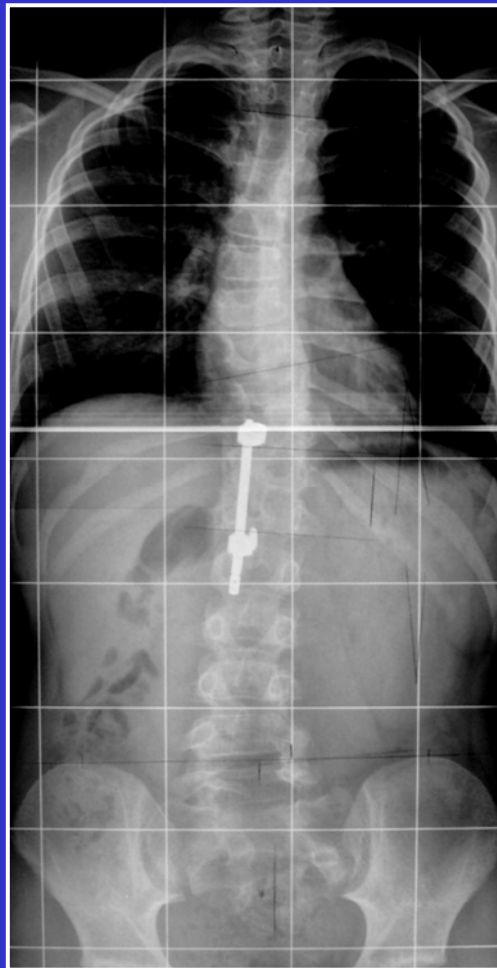
13A 10M



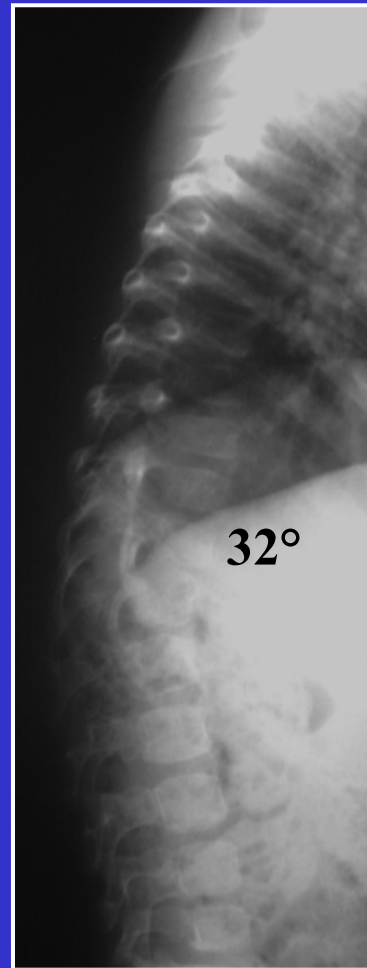
GT 5 mm  
TT 5 mm



1A 2M



9A 4M



1A 2M



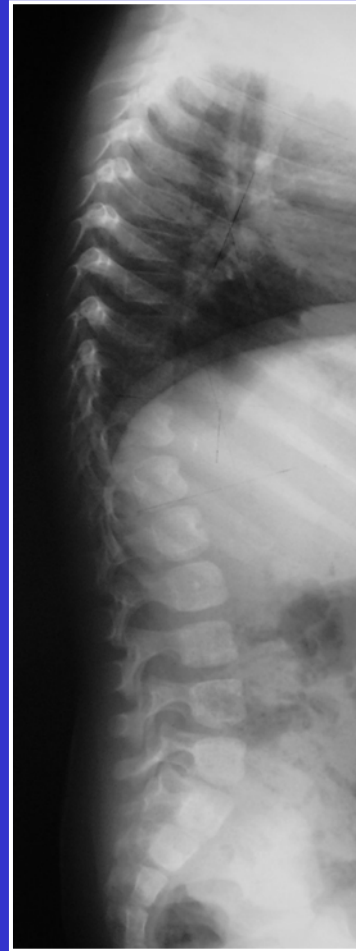
9A 4M



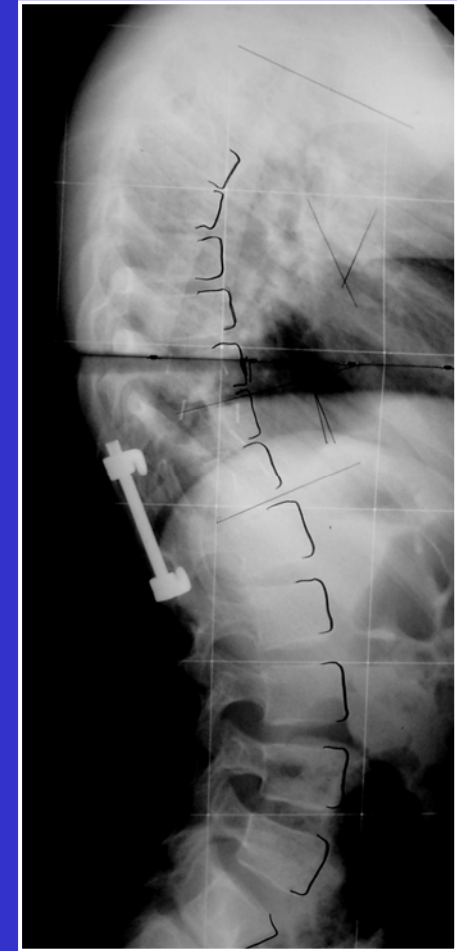
1A 6M



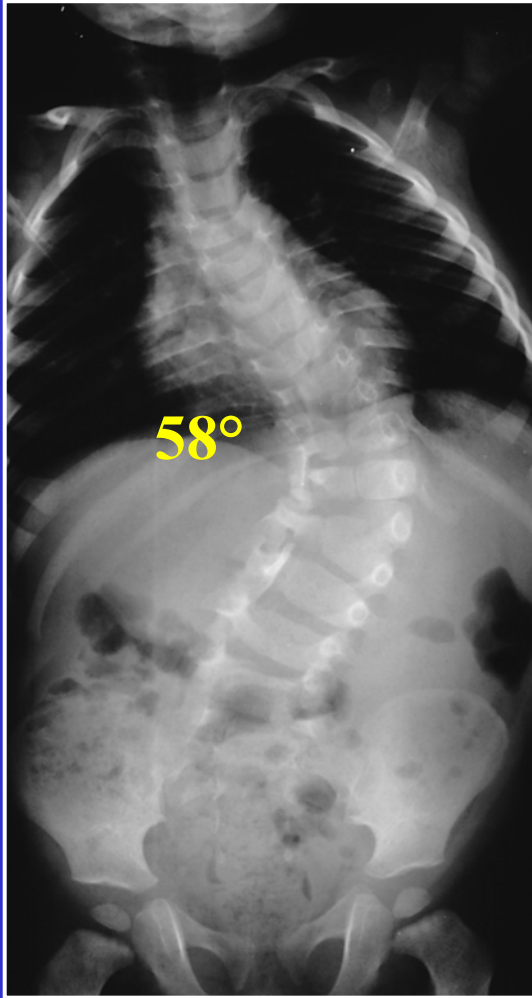
8A 4M



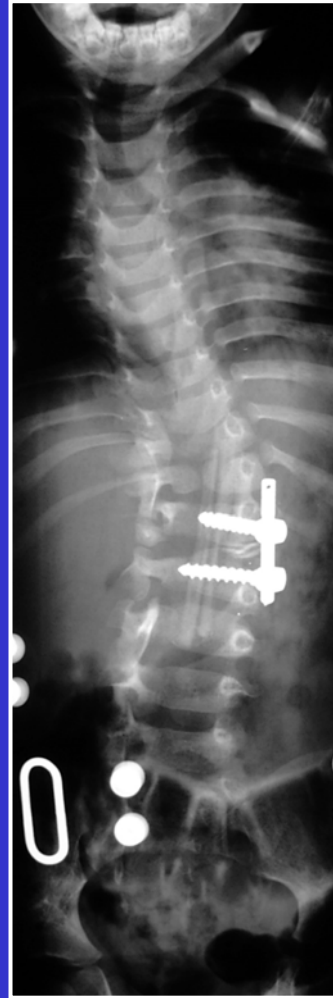
1A 6M



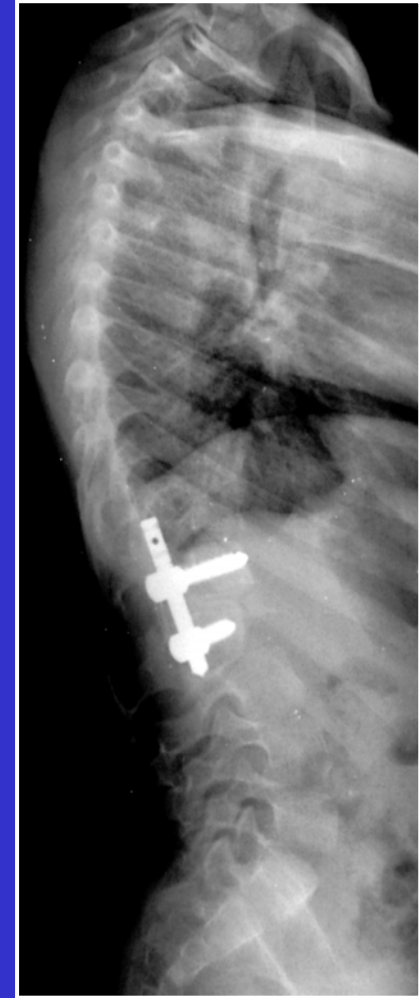
8A 4M



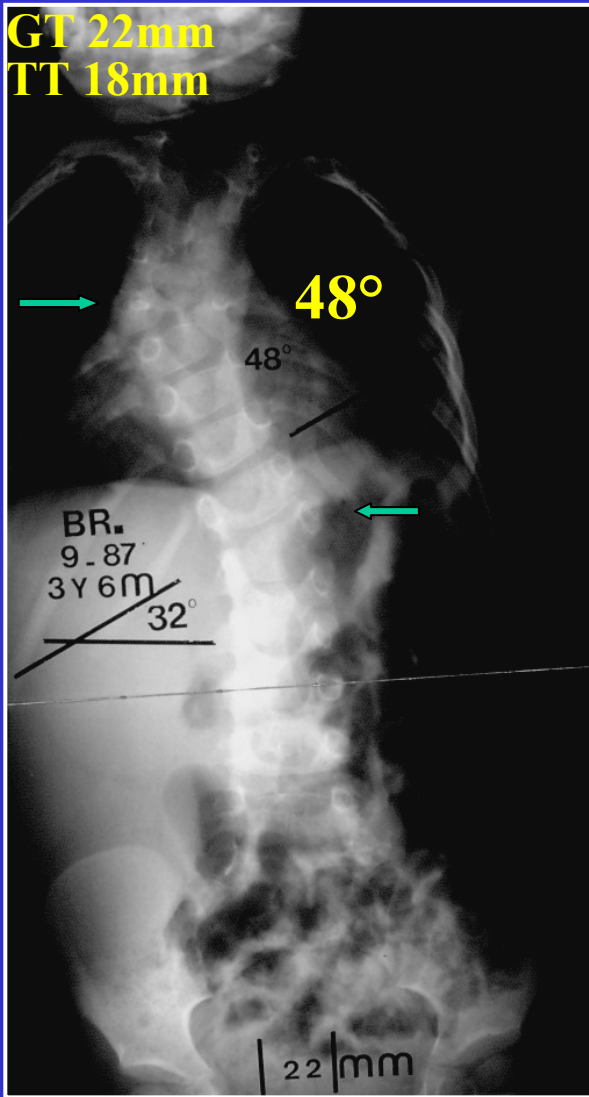
**1A 9M**



**J + 1M**



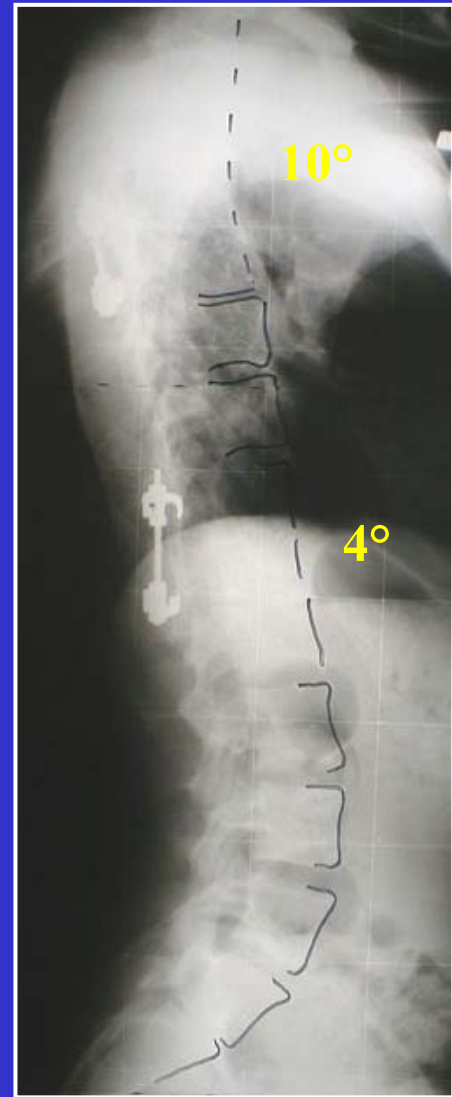
**J+1M**



3 A 6 M



17 A



17 A

## 26 HEMIVERTEBRES LOMBAIRES

LOCALISATION ENTRE L2 ET L4

SEXE : 11 FILLES 15 GARCONS

6 DROITES 20 GAUCHES

SEGMENTATION { 14 COMPLETEMENT SEGMENTEES  
12 SEMI SEGMENTEES

AGE à L'OPERATION 3.4 A (12M – 10.2A)

SUIVI MAX. 6A 9M (6M – 17.4A)

DUREE OPERATOIRE 302' (240' – 450')

PERTE D'HEMOGLOBINE 3.3 g/dl (1.9 – 5.9)



## **Malformations congénitales associées**

**VATER, imperforation anale, atrésie de l'oesophage, craniosténose, reflux vésico-urétéral droit, malformation du pouce**

**agénésie rénale**

**atrésie de l'oesophage, trachéomalacie, diverticule de Meckel, omphalocèle**

**agénésie du pouce gauche, pouce droit tri phalangien.**

**reflux vésico-urétéral, agénésie sacrée distale.**

**agénésie rénale, hypospadias, hypoplasie du pouce droit.**

**imperforation anale, exstrophie vésicale.**

**Atrésie de l'oesophage avec fistule oeso trachéale.**

**Reflux vésico-urétéral gauche, rein droit polykystique (exérèse), veine cave supérieure se drainant dans le sinus coronaire.**

**reflux vésico-urétéral bilatéral.**

## **Malformations médullaires associées.**

**Moelle fixée avec lipome, syringomyélie.**

**Moelle fixée.**

**Amputation distale du cône terminal.**

**Syringomyélie.**

**Chirurgies additionnelles.**

**Arthrodèse postérieure avec mini Harrington pour cyphose évolutive.**

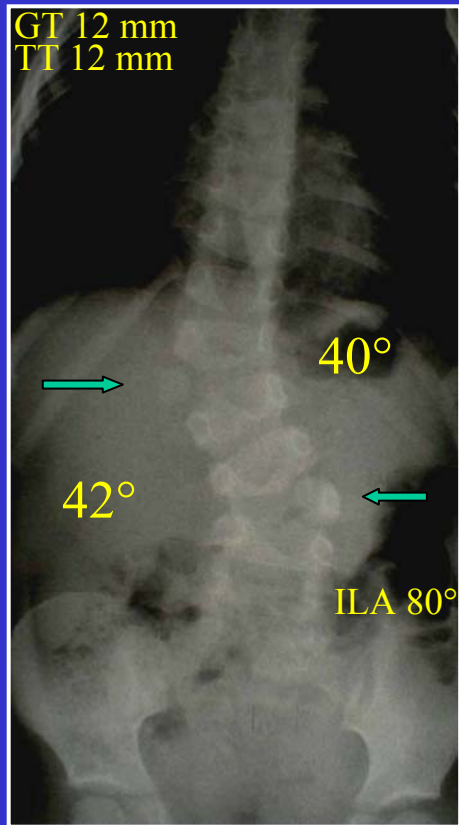
**Changement de matériel pour fracture de tige.**

**Changement de crochet.**

<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	32.6°(24-46)	10.8° (2-24)	8.76° (0-28)
Courbure principale globale (°)	33.5° (24-46)	12.6° (2-26)	10.9° (0-36)
Contre courbure craniale (°)	16.5° (4-42)	6.9° (0-34)	6.5° (0-29)
Contre courbure caudale (°)	7.2° (0-32)	2.7° (0-11)	1.3° (0-12)
Translation gravitaire (mm)	11.4 <sub>mm</sub> (1-34)	12.4 <sub>mm</sub> (0-44)	9.3 <sub>mm</sub> (0-30)
Translation gravitaire (%)	11%(1-32)	11% (0-35)	7% (0-21)
Translation vraie (mm)	18.3 <sub>mm</sub> (1-59)	17.9 <sub>mm</sub> (2-44)	11.8 <sub>mm</sub> (0-57)
Translation vraie (%)	18% (1-48)	17% (2-43)	9%(0-40)

<b>LORDOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	-2.6°(-24/22)	-4.9°(-28/22)	-5.7° (-28/34)
Courbure principale globale (°)	-26.3°(-56/10)	-21.7°(-44/34)	-31.7°(-60/30)

<b>LORDOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	-2.6°(-24/22)	-4.9°(-28/22)	-5.7° (-28/34)
Courbure principale globale (°)	-26.3°(-56/10)	-21.7°(-44/34)	-31.7°(-60/30)



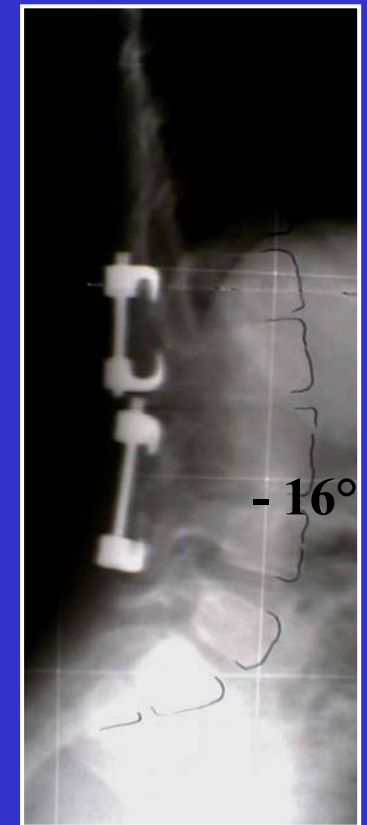
5 A



6 A



5 A



6 A

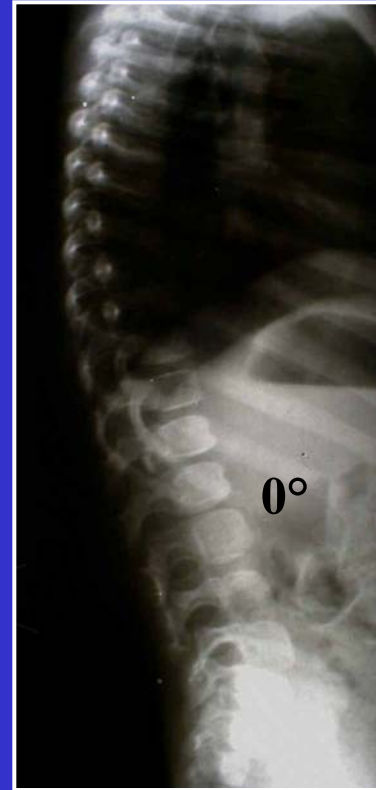




**1 A**



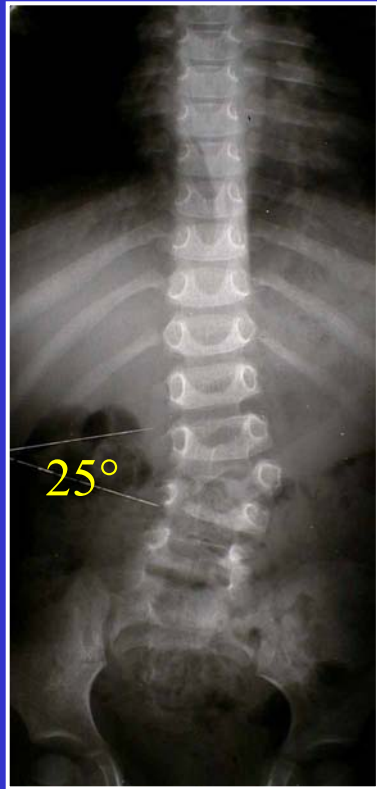
**13 A 5 M**



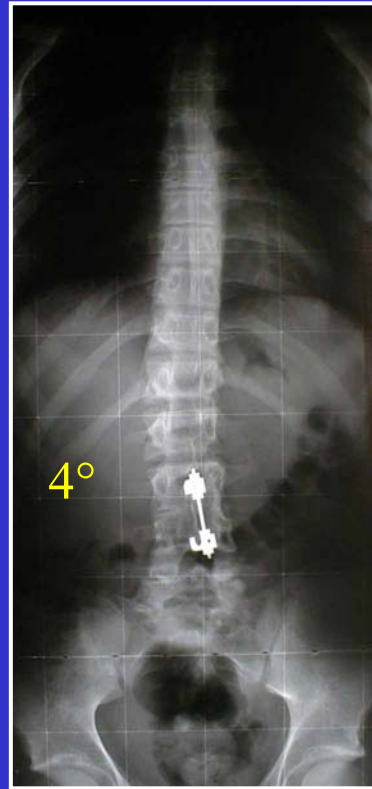
**1 A**



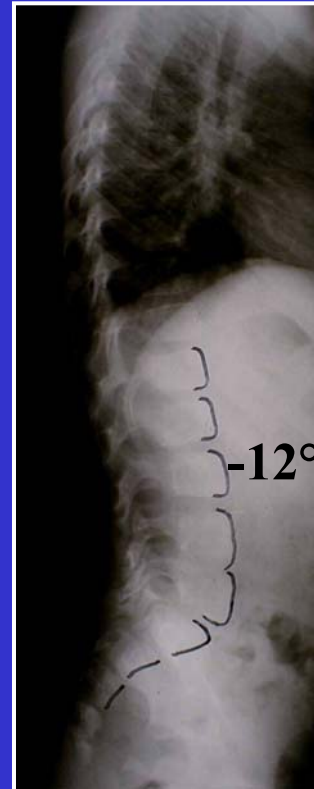
**13 A 5 M**



**2 A 11 M**



**15 A 1 M**



**2 A 11 M**



**15 A 1 M**



**8 M**



**18 A 5 M**



**8 M**



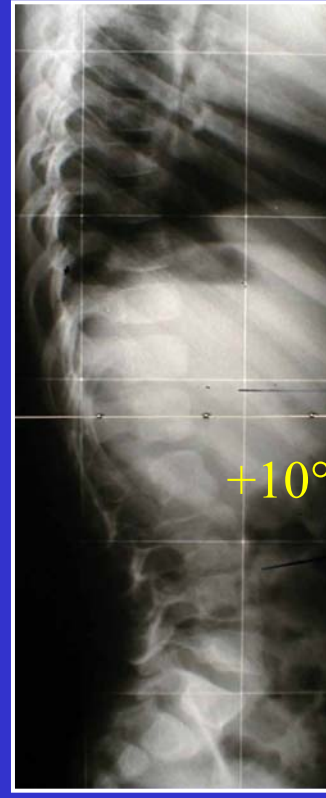
**18 A 5 M**



**4 A 11 M**



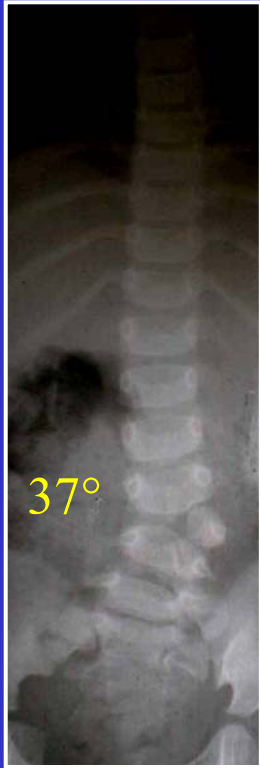
**13 A 5 M**



**4 A 11 M**



**13 A 5 M**



**1 A 8 M**



**13 A 10 M**



**1 A 8 M**



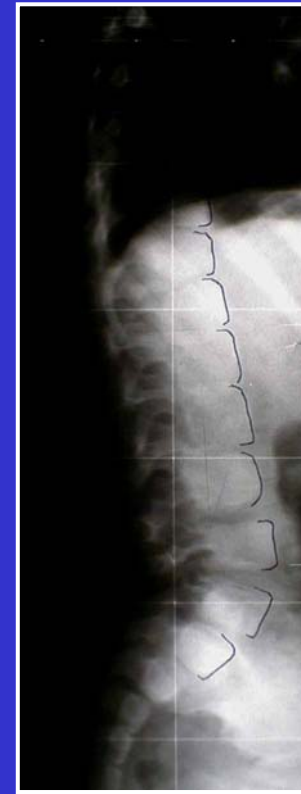
**13 A 10 M**



**4 A 8 M**

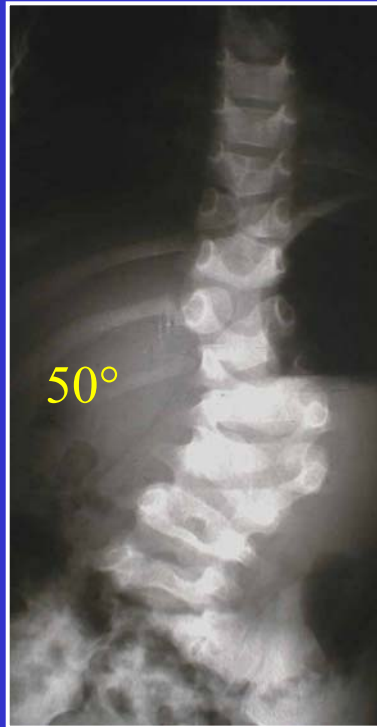


**5 A 8 M**



**4 A 8 M**

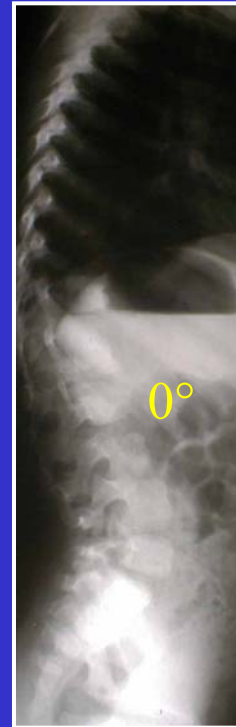




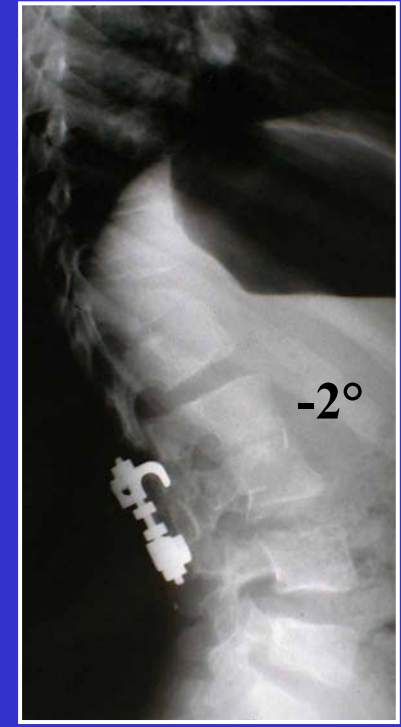
**3 A 6 M**



**9 A 1 M**



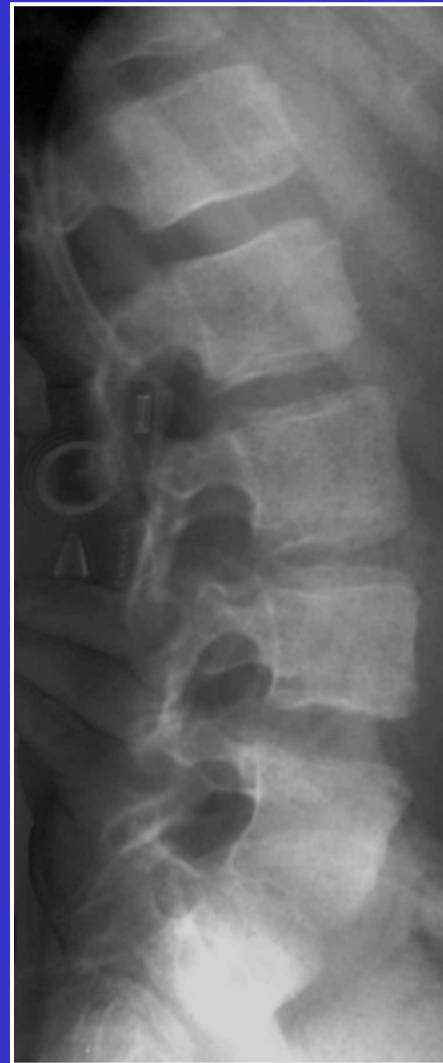
**3 A 6 M**

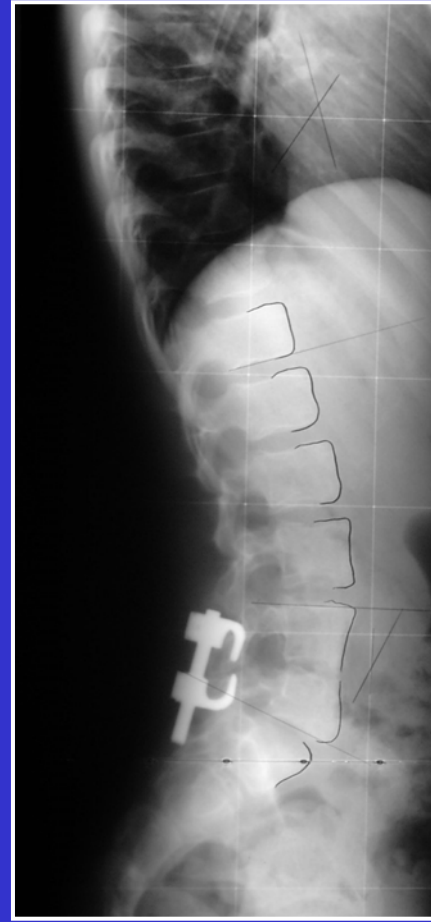
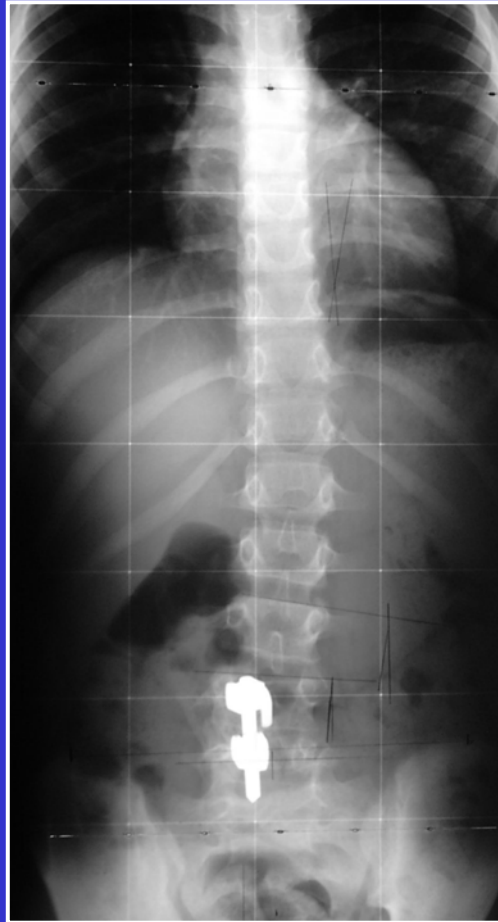


**9 A 1 M**



**3. 2002**  
**9 A 10 M**





**7. 2004 12 A 2 M**

## 17 HEMIVERTEBRES LOMBO - SACREES

LOCALISATION ENTRE L4 ET S1

10 DROITES 7 GAUCHES

SEXE: 9 FILLES 8 GARCONS

SEGMENTATION { 10 COMPLETEMENT SEGMENTEES  
7 SEMI SEGMENTEES

AGE à L'OPERATION 4.9 A (3M – 10.3A)

SUIVI MAX. 7.1 A (6M – 17.8A)

DUREE OPERATOIRE 290' (240' – 345')

PERTE D'HEMOGLOBINE 3.7 g/dl (1.4 – 7)

4 patients transfusés

## **Malformations médullaires associées.**

**Moelle fixée avec lipome.**

**Méningocèle.**

**Myélocystocèle.**

**Méningocèle et moelle fixée avec lipome.**

## **Chirurgie additionnelle.**

**Ablation de matériel.**

**Ablation de matériel, tige sous cutanée sans arthrodèse, 3 allongements de tige.**

**Arthrodèse convexe itérative avec mini Harrington pour pseudarthrose, ablation de matériel.**



<b>SCOLIOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	26.9°(13-42)	10.4° (2-18)	9.1° (2-17)
Courbure principale globale (°)	27.8°(13-42)	12.3° (2-20)	11.4° (2-20)
Contre courbure craniale (°)	25.7° (13-44)	11.7° (4-22)	12.5° (0-22)
Translation gravitaire (mm)	16.6 <sub>mm</sub> (0-38)	12.6 <sub>mm</sub> (0-35)	14.5 <sub>mm</sub> (1-40)
Translation gravitaire (%)	15% (0-34)	11% (0-30)	11% (1-40)
Translation vraie (mm)	19.2 <sub>mm</sub> (2-43)	19.4 <sub>mm</sub> (2-67)	25.5 <sub>mm</sub> (2-59)
Translation vraie (%)	18% (2-49)	16% (2-61)	17% (3-43)

<b>LORDOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	-13.5°(-2/-26)	-14.5°(-6/-30)	-12° (0/-24)
Courbure principale globale (°)	-36°(-14/-54)	-34°(-16/-51)	-43.7°(-9/-67)

<b>LORDOSES</b>	<b>PRE-OP</b>	<b>POST-OP</b>	<b>ReculMAX</b>
Courbure segmentaire (°)	-13.5°(-2/-26)	-14.5°(-6/-30)	-12° (0/-24)
Courbure principale globale (°)	-36°(-14/-54)	-34°(-16/-51)	-43.7°(-9/-67)



**9A 4 M**



**14 A 4 M**



**9 A 4 M**



**14 A 4 M**



**5 A 10 M**



**16 A 4 M**



**5 A 10 M**



**16 A 4 M**



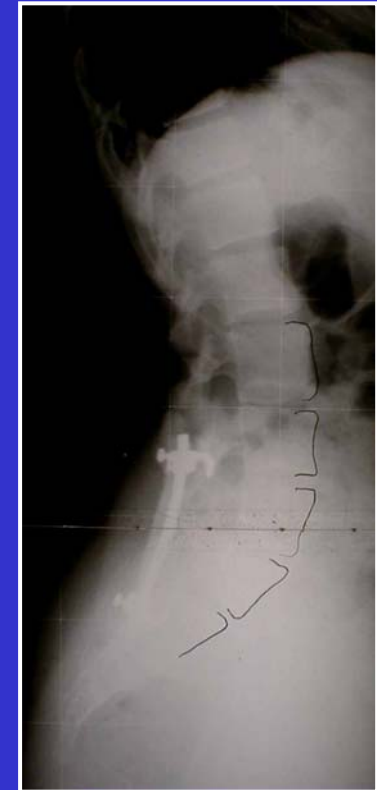
**10 A 4 M**



**15 A 7 M**



**10 A 4 M**



**15 A 7 M**



**4 A 10 M**



**9 A 4 M**



**9 A 4 M**



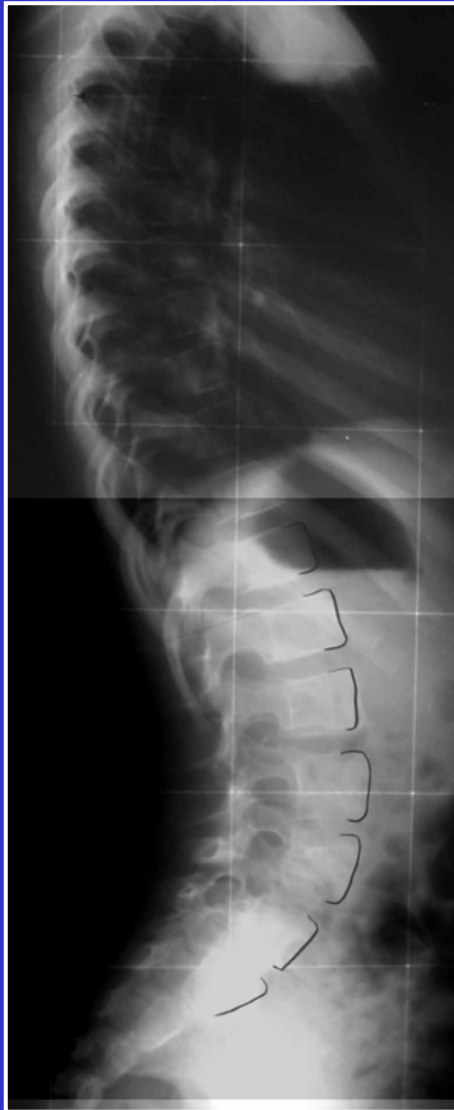
**5 A 11 M**



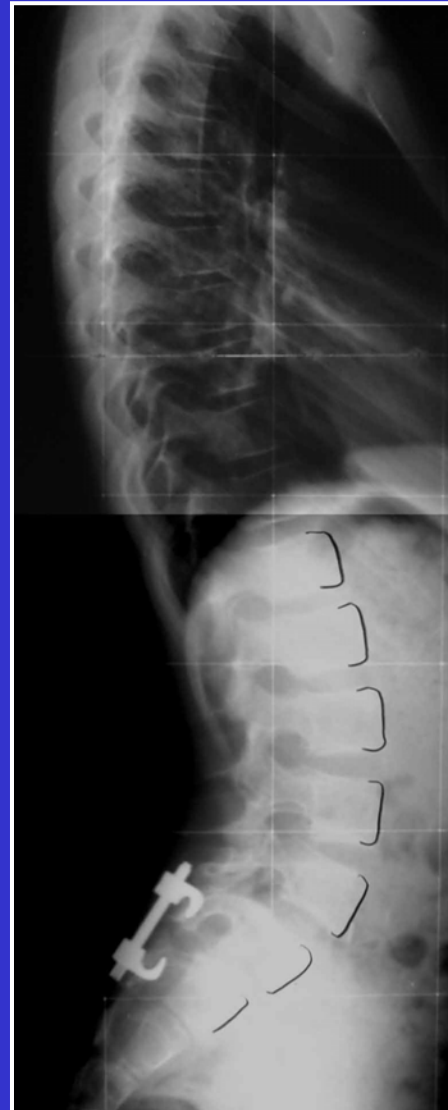
**8 A 10 M**

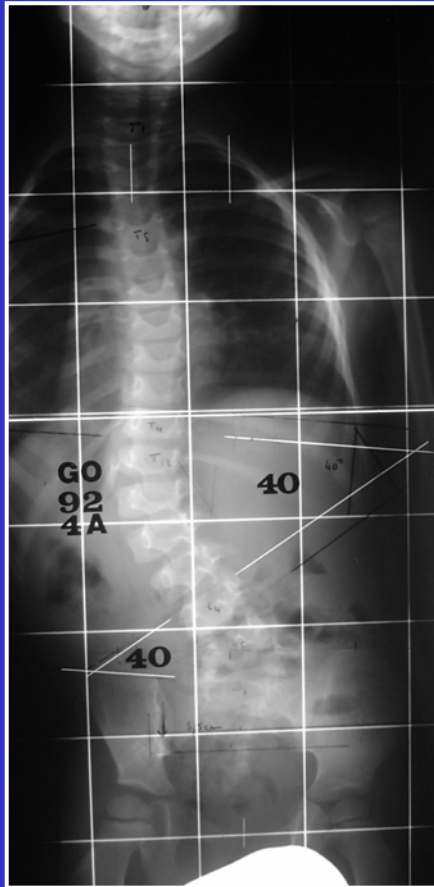


**5 A 11 M**

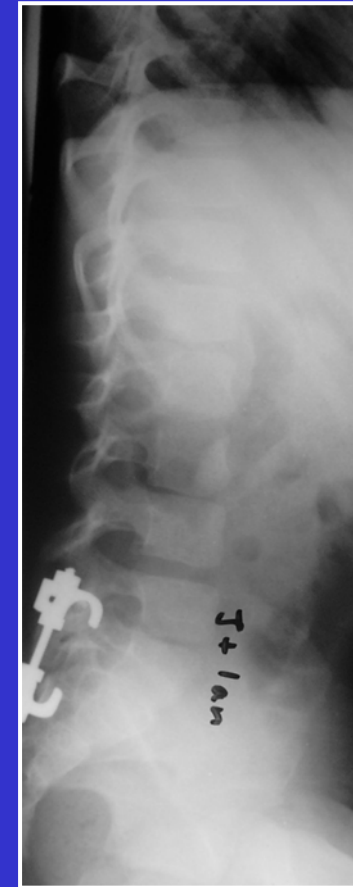
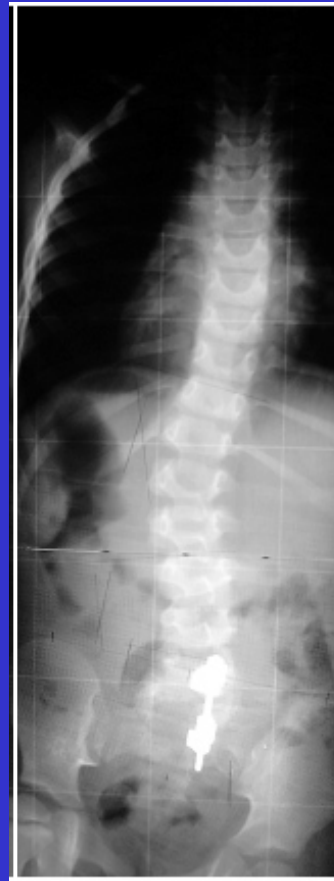


**8 A 10 M**





4 A 3 M



6 A 9 M

**HEMI-VERTEBRE C7 / T1**

**2+1/2 A**

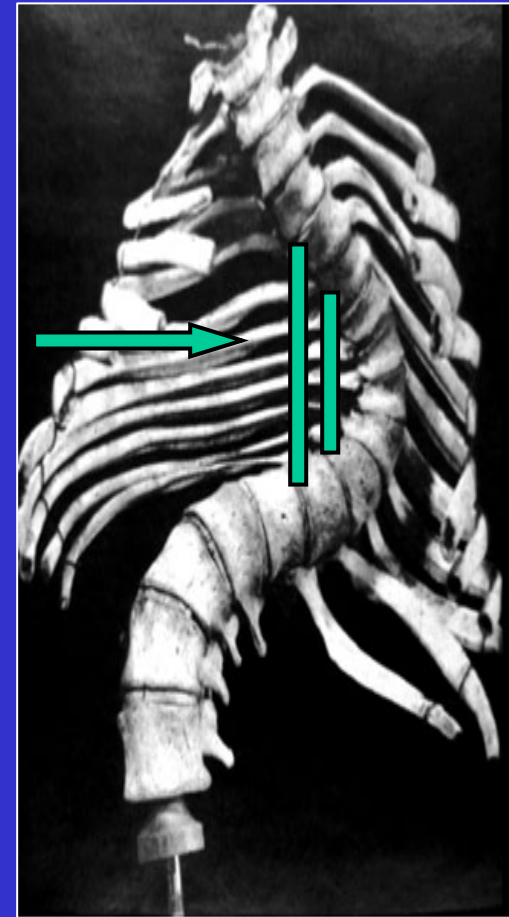
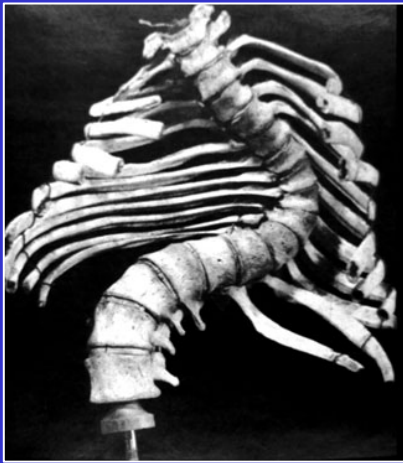


**5 A**



1

TRACTION PAR HALO-FAUTEUIL OU HALO-VESTE SI APEX REDUCTIBLE, SINON LE LIBERER

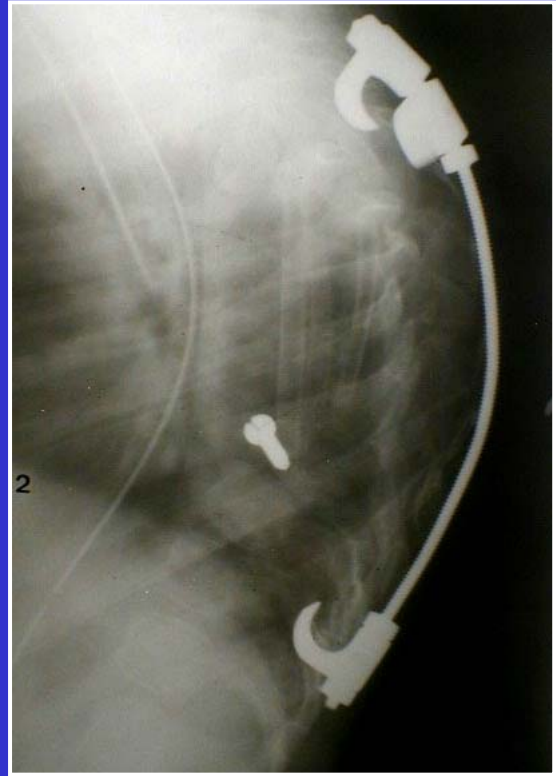


3

PUIS GREFFE ET INSTRUMENTATION POSTERIEURE

2

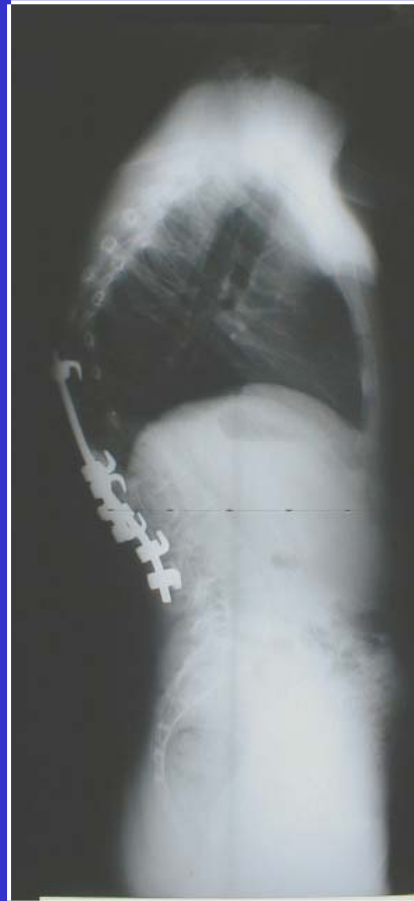
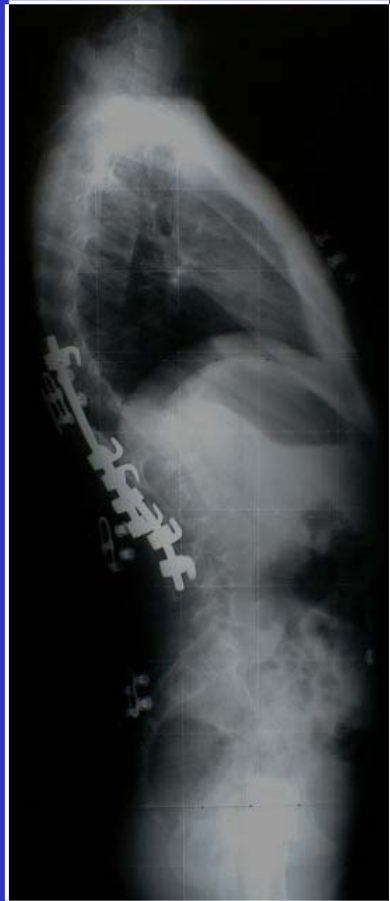
PUIS GREFFE ANTERIEURE PAR LA CONCAVITE











## **CONCLUSION 1**

**La constatation d'une malformation congénitale du rachis implique une recherche de malformations associées**

## **CONCLUSION 2**

**Toutes les scoliozes congénitales ne sont pas évolutives et ne nécessitent donc pas forcément de prise en charge**

## **CONCLUSION 3**

**Lorsque la scoliose congénitale est évolutive le traitement chirurgical est indiqué idéalement avant l'âge de trois ans**

#### **CONCLUSION 4**

L'arthrodèse convexe reste une indication de choix pour les localisations thoraciques d'H.V. et/ou les puzzles vertébraux

#### **CONCLUSION 5**

La résection d'une hémivertèbre thoraco-lombaire, lombaire ou lombo-sacrée est une chirurgie aux risques contrôlés sans complication majeure dans notre série.

#### **CONCLUSION 7**

Dans les formes opérées tardivement libération, traction, arthrodèse postérieure et antérieure concave sont indiquées

#### **CONCLUSION 8**

Toutes ces conditions et en particulier la précédente s'accompagne d'un risque neurologique non négligeable



