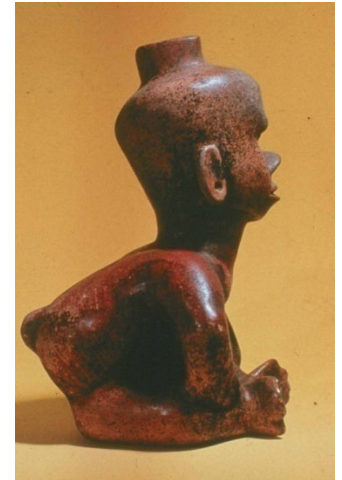


# Congenital dislocation of the spine and Congenital Kyphosis Type II



G BOLLINI



Timone Children's hospital



Marseilles

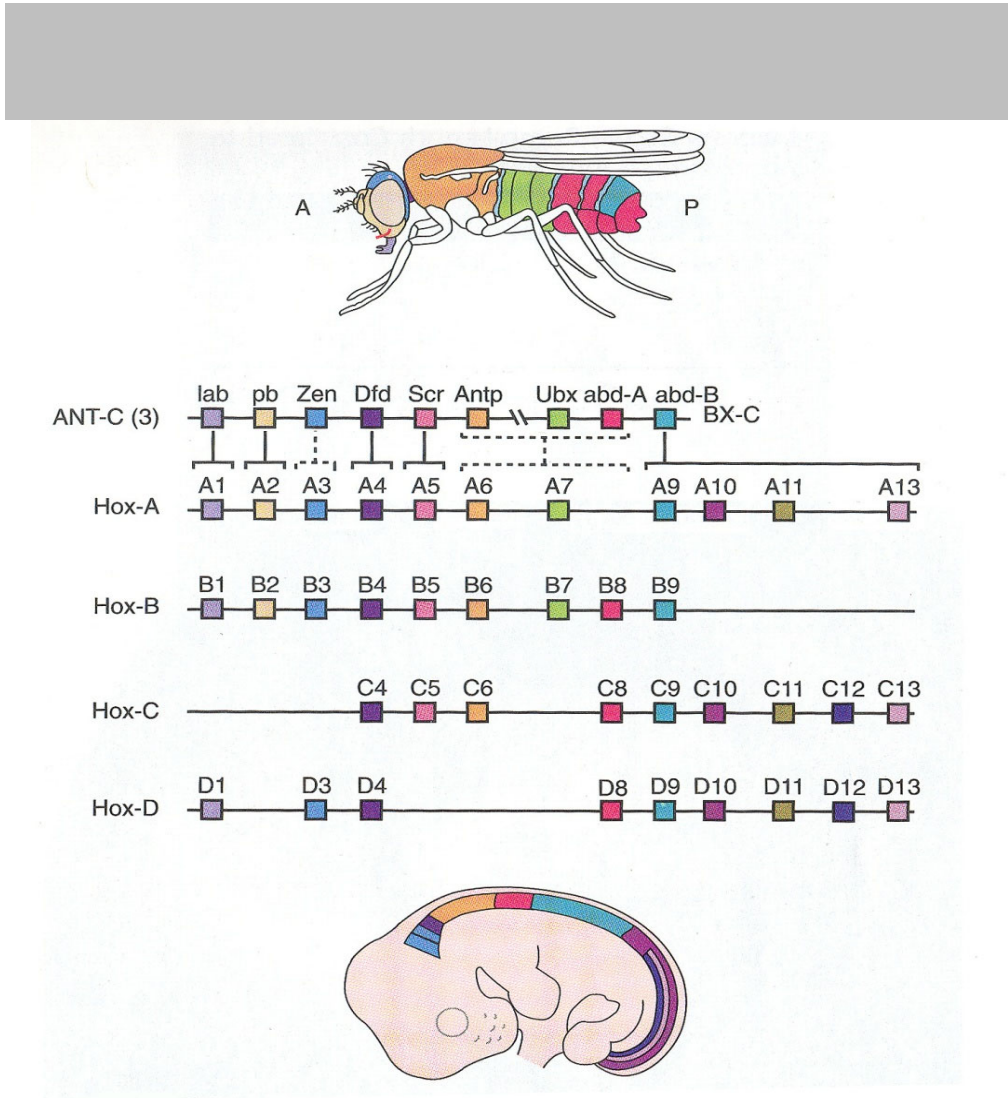
DISCLOSURE

Consultant:

SYNTHES

MEDTRONIC

Malformations are correlated with a dysfunction of the genes coding for segmentation



Colour correlation between the segmentation genes and the somites

## WINTER CLASSIFICATION

DEFECT of FORMATION

TYPE I

**ANTERO-LATERAL DEFECT**

ANTERIOR DEFECT

DEFECT of SEGMENTATION

TYPE II

COMPLETE ANTERIOR FUSION

PROGRESSIVE ANTERIOR FUSION

COMPLEXE MALFORMATION

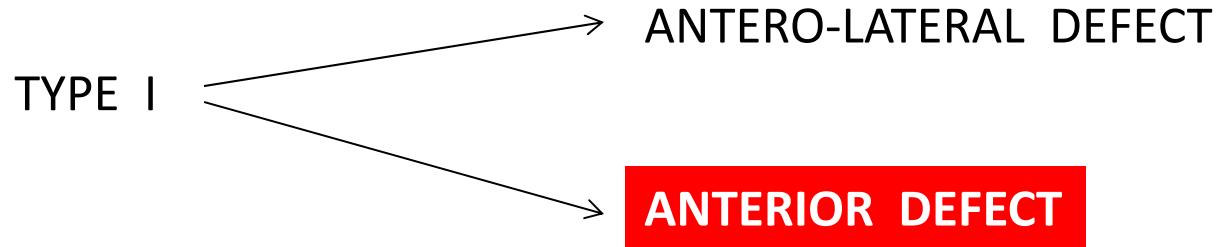
TYPE III

## ANTERO-LATERAL DEFECT

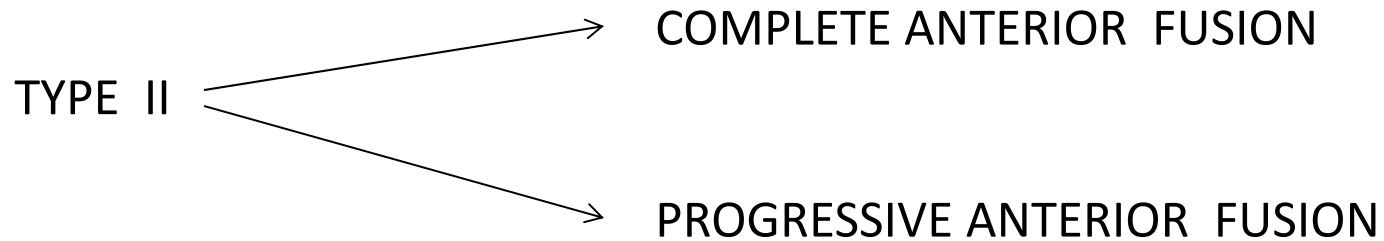


## WINTER CLASSIFICATION

DEFECT of FORMATION



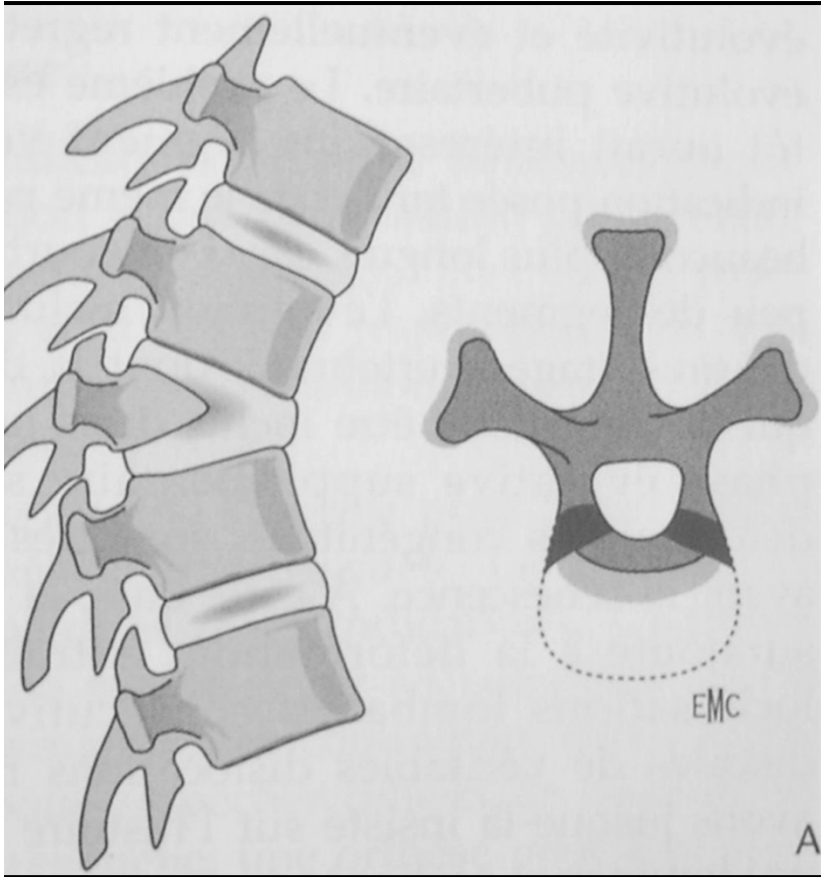
DEFECT of SEGMENTATION



COMPLEXE MALFORMATION

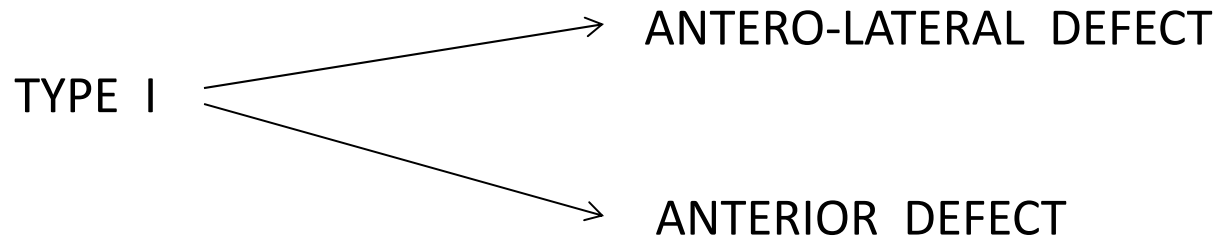
TYPE III

# ANTERIOR DEFECT



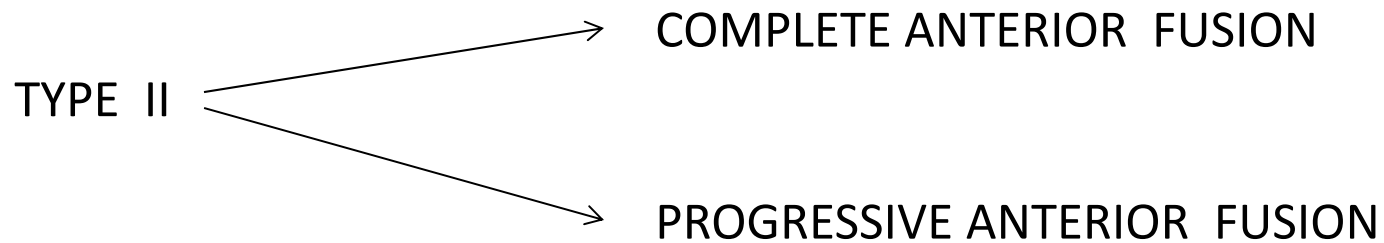
## MODIFIED WINTER CLASSIFICATION

DEFECT of FORMATION



**PEDICULAR DEFECT, POSTERIOR DEFECT and FULL SEGMENTAL DEFECT**

DEFECT of SEGMENTATION



COMPLEXE MALFORMATION

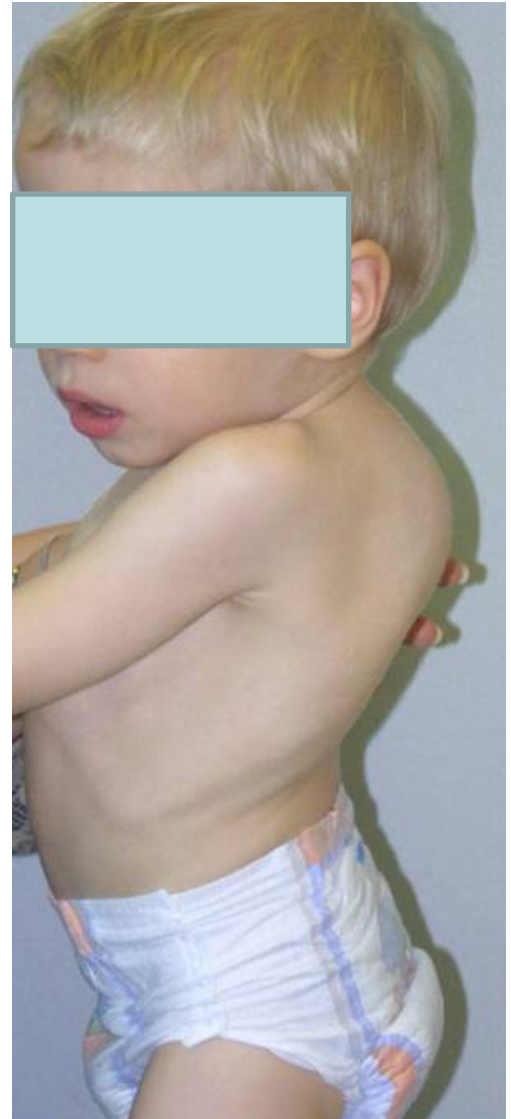
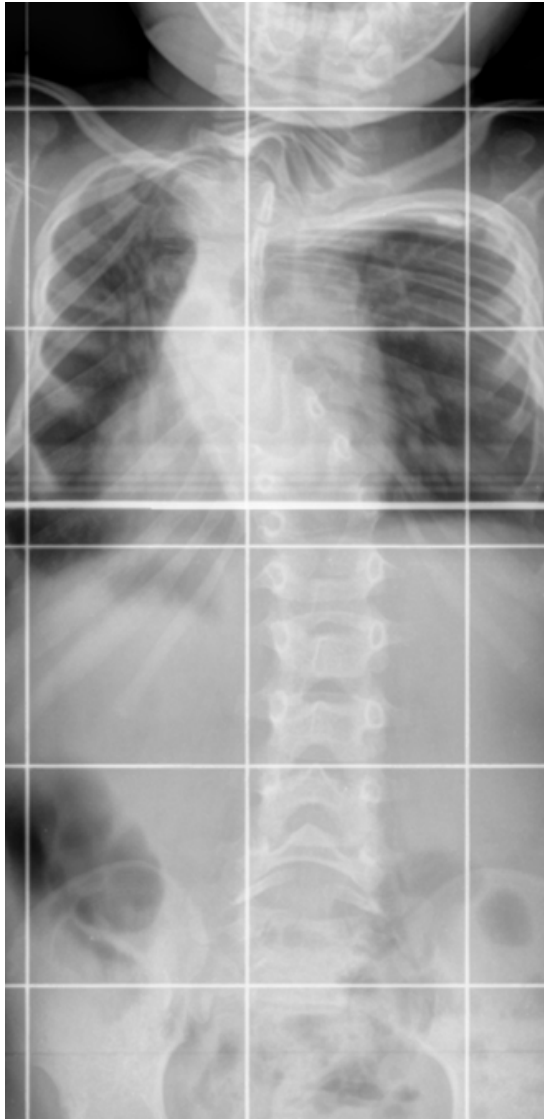
TYPE III



**PEDICULAR DEFECT**



**PEDICULAR DEFECT**

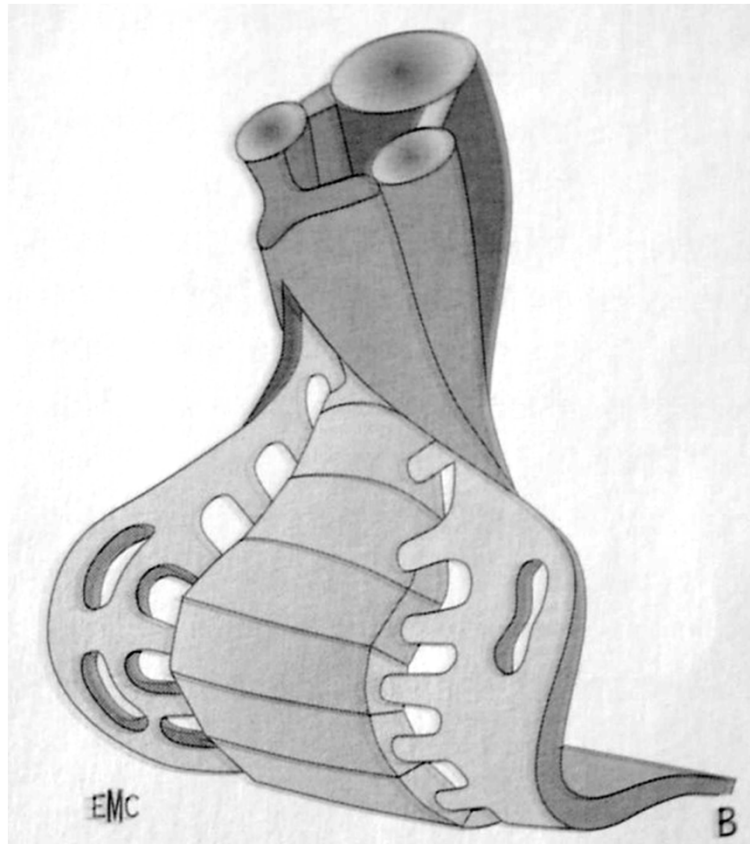


2 Y

# PEDICULAR DEFECT



**POSTERIOR DEFECT**



MYELOMENINGOCELE  
MIXED CONGENITAL AND NEUROLOGIC KYPHOSIS

**FULL SEGMENTAL DEFECT**

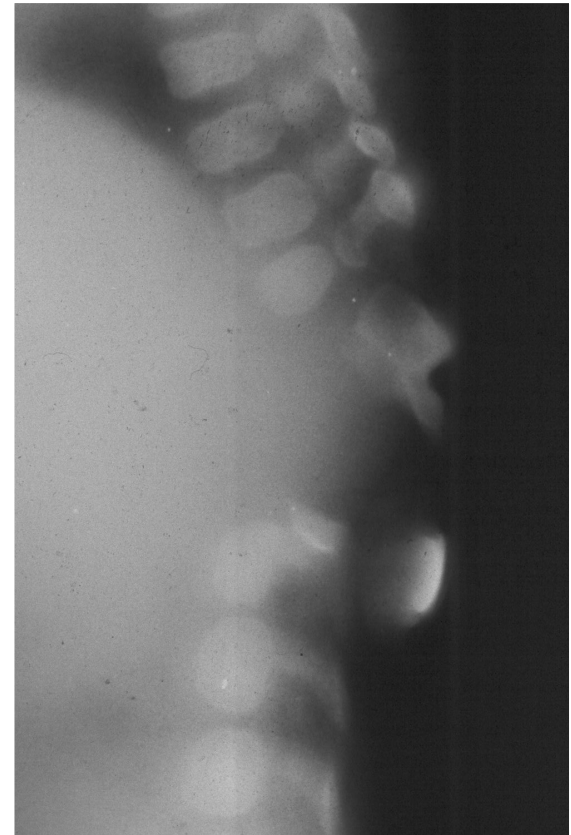
**TERMINAL**



**LUMBO-SACRAL AGENESIA**

**FULL SEGMENTAL DEFECT**

**INTERCALARY**



FULL SEGMENTAL DEFECT

INTERCALARY

CONGENITAL SPINE DISLOCATION



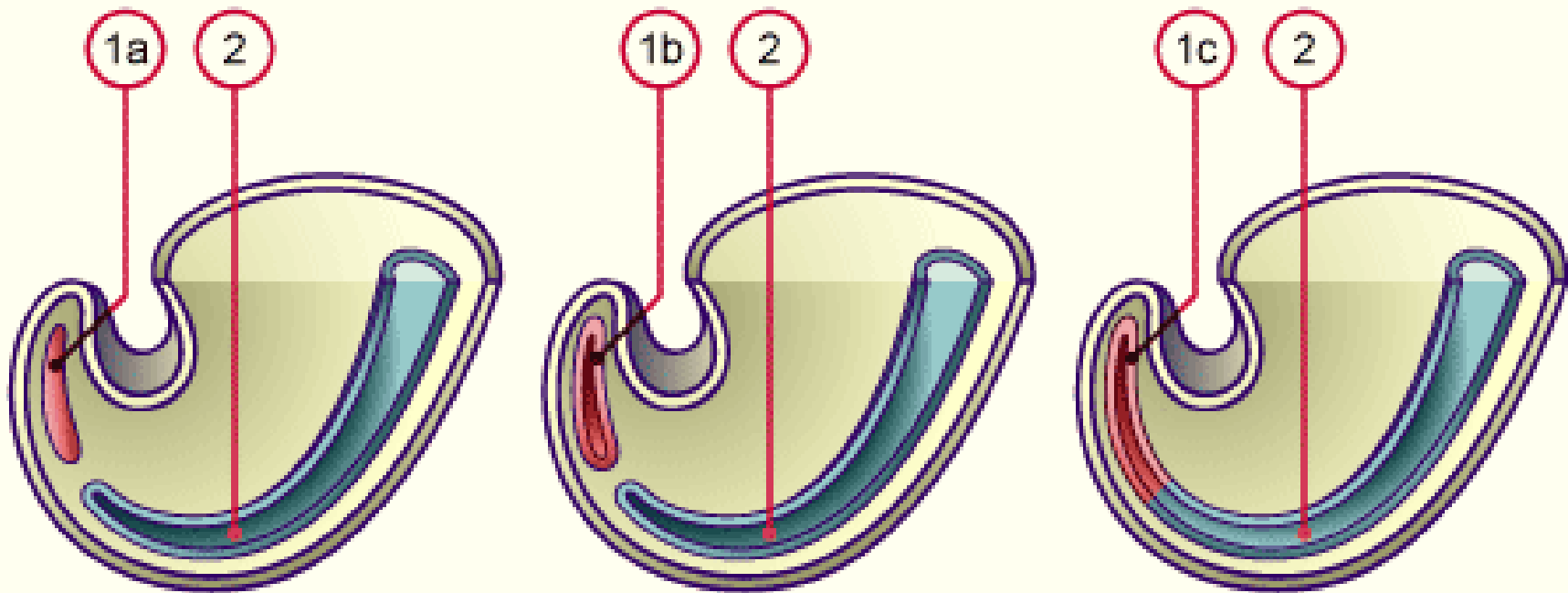
### Congenital Dislocated Spine

Elke Viehweger, MD, PhD,\* Marie-Christine Giacomelli, MD,† Yann Glard, MD,\*

Franck Launay, MD, PhD,\* Philippe Petit, MD,† Jean-Luc Jouve, MD, PhD,\* and Gerard Bollini, MD\*

J Pediatr Orthop 2009;29:362Y368

## CONGENITAL SPINE DISLOCATION



After the neural tube has closed, the caudal extremity keeps developing beyond

The caudal cord/spine develop under the ectoderm before regressing partially (in primates).



## CONGENITAL SPINE DISLOCATION

EARLY STABILIZATION IS MANDATORY



BIRTH



9 Y

**CONGENITAL SPINE DISLOCATION**

2+6 Y



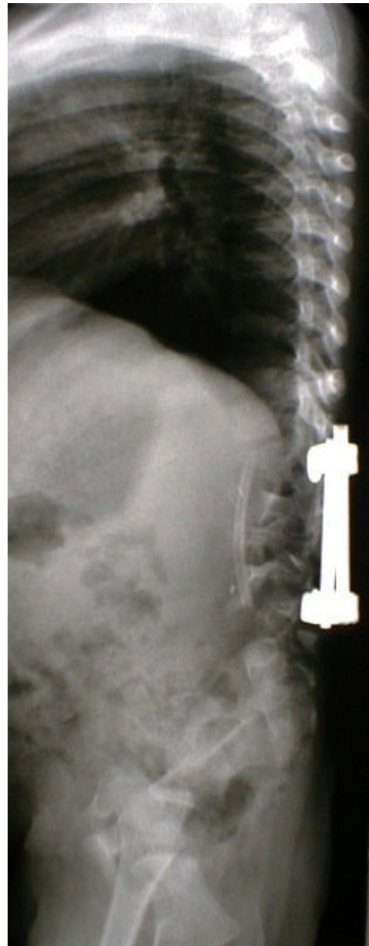
13 Y



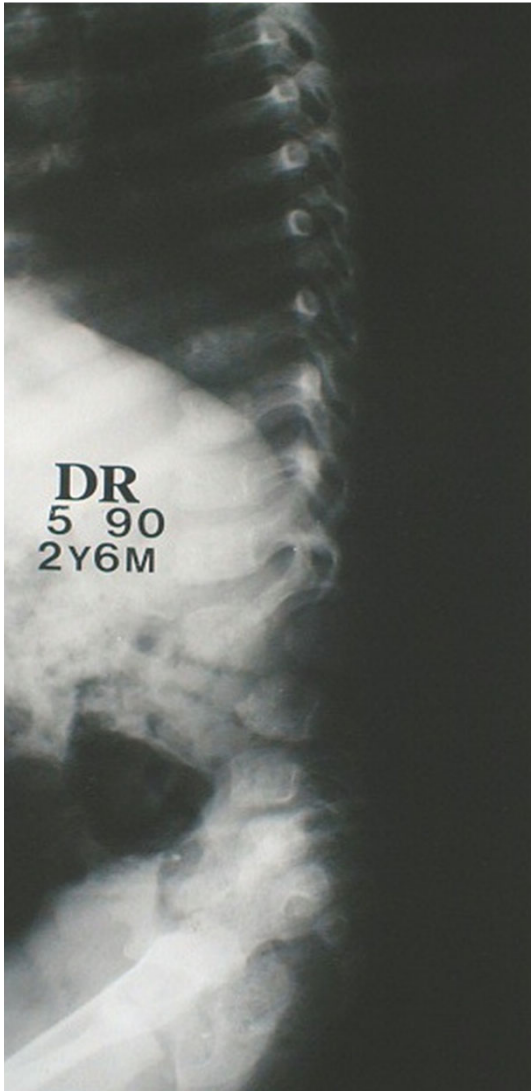
# CONGENITAL SPINE DISLOCATION



# CONGENITAL SPINE DISLOCATION

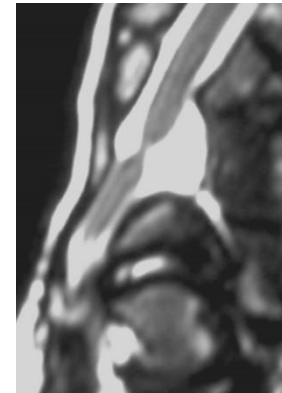


**CONGENITAL SPINE DISLOCATION**



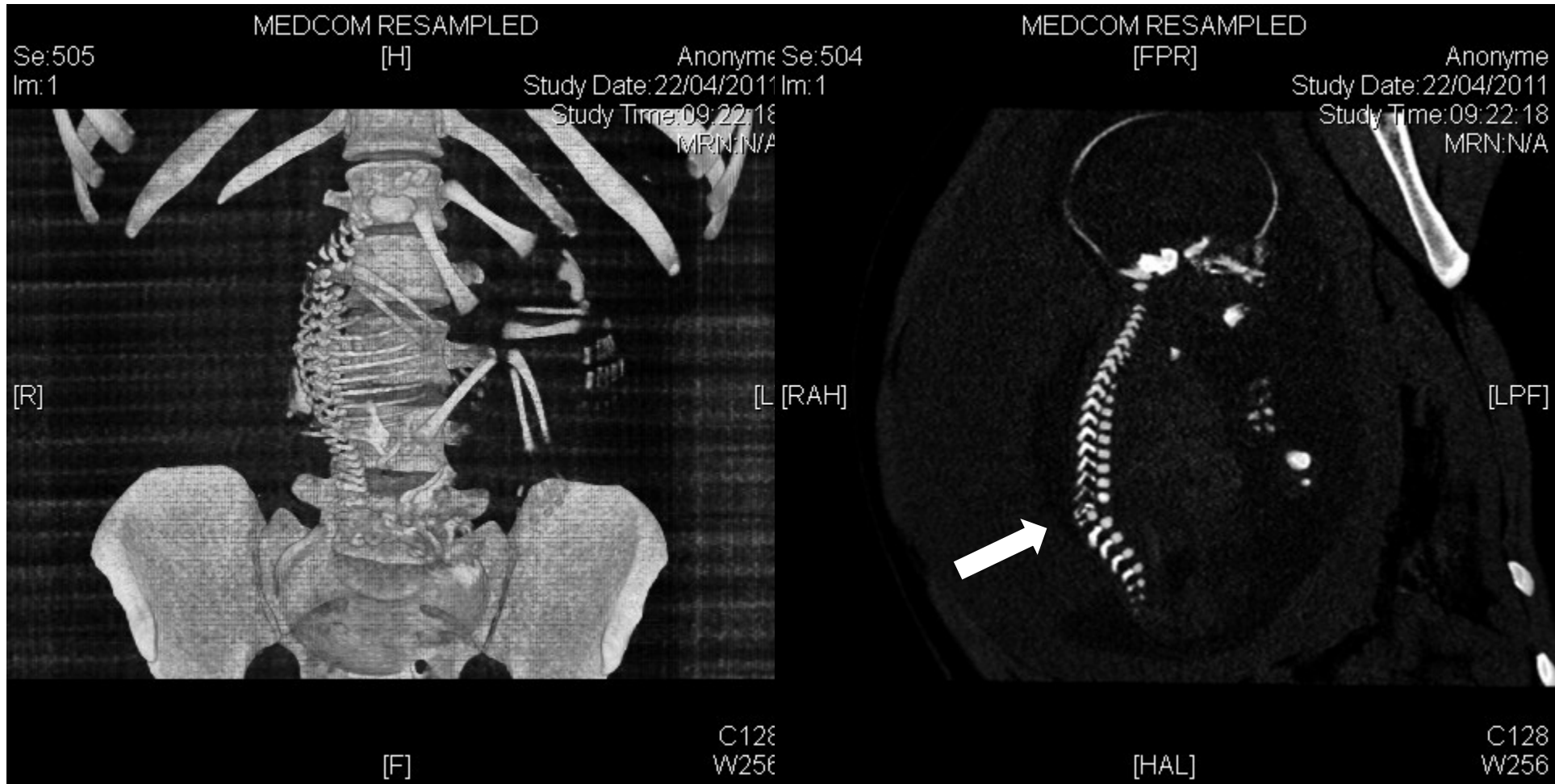
## CONGENITAL SPINE DISLOCATION

Neurologic status extends from total paraplegia to mild paraparesia



Associated Regional  
Cord Dysplasia

# CONGENITAL SPINE DISLOCATION



PRE-NATAL DIAGNOSIS

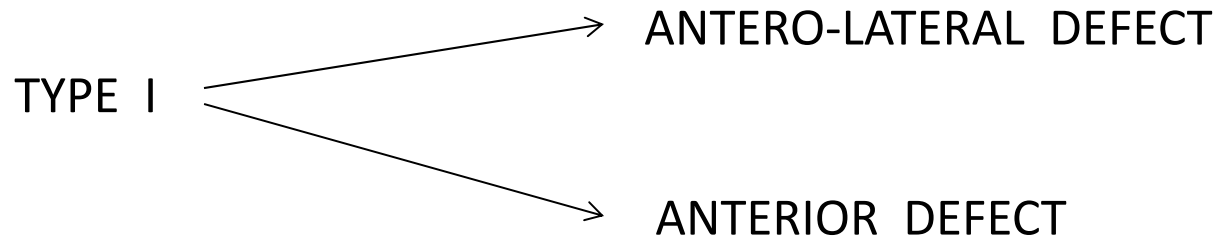
# CONGENITAL SPINE DISLOCATION





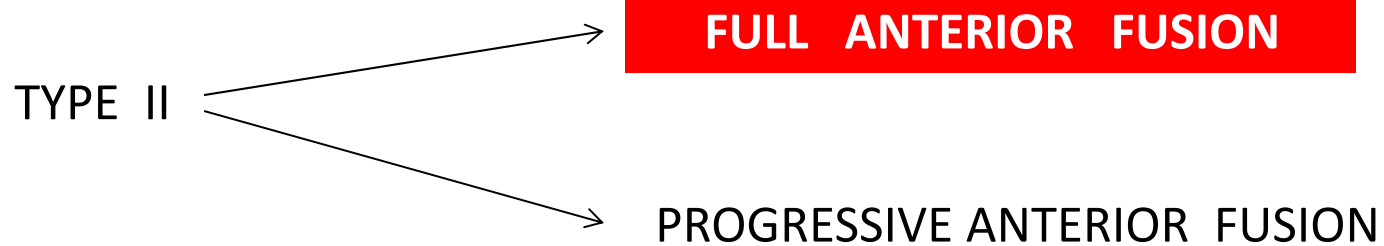
# MODIFIED WINTER CLASSIFICATION

DEFECT of FORMATION



PEDICULAR DEFECT, POSTERIOR DEFECT and FULL SEGMENTAL DEFECT

DEFECT of SEGMENTATION



COMPLEXE MALFORMATION

TYPE III

**FULL ANTERIOR FUSION**

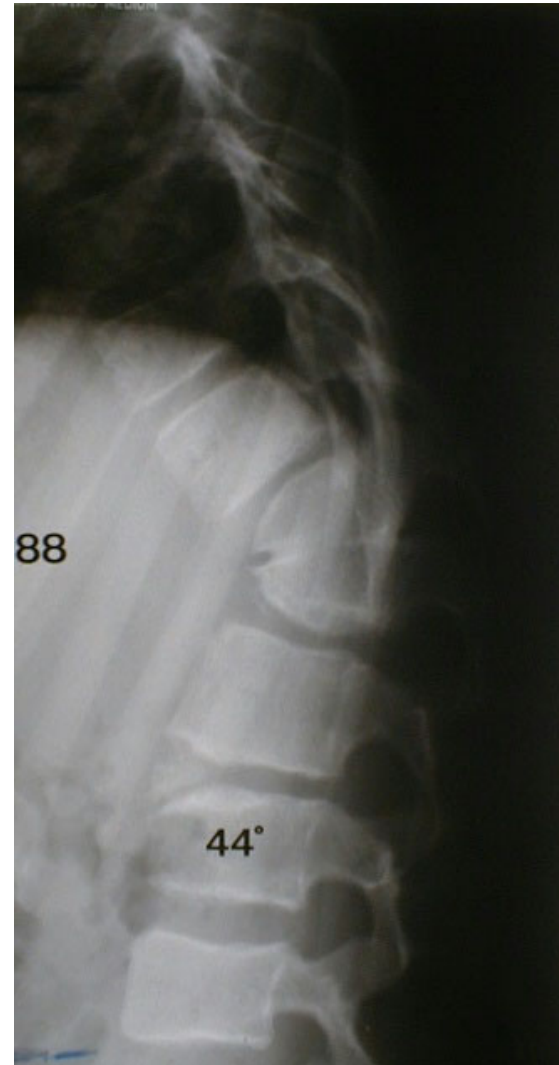
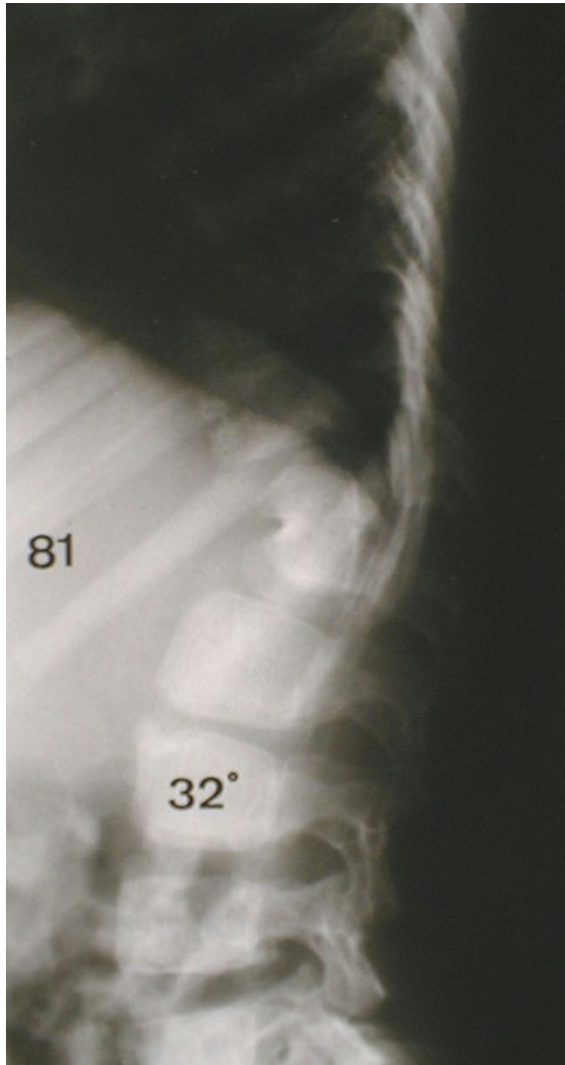


MILD

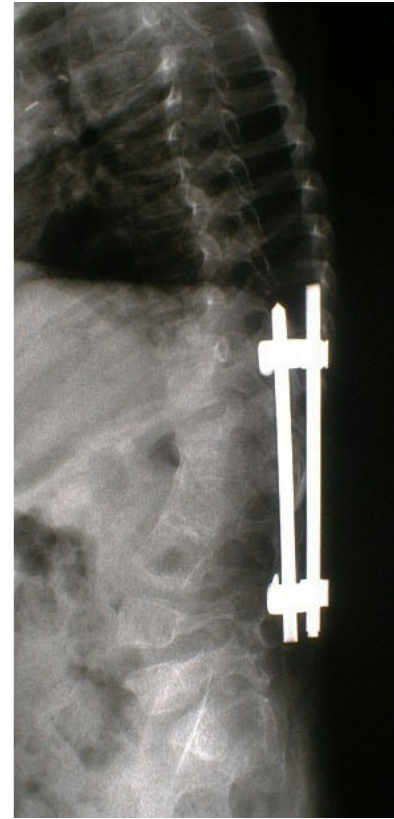
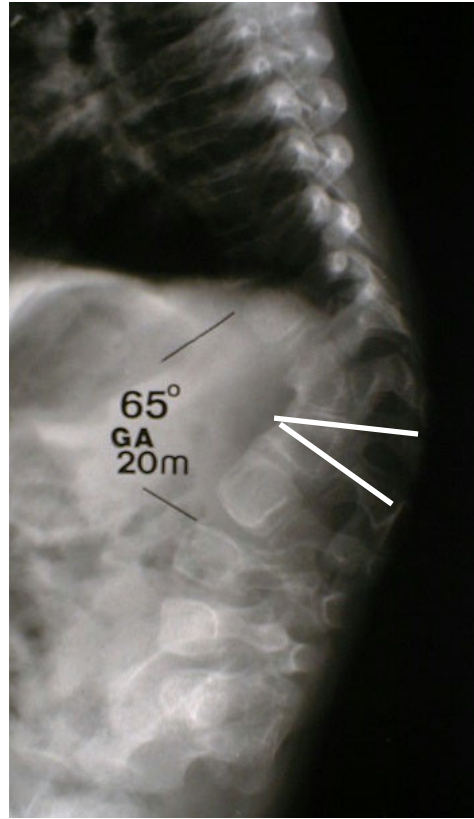
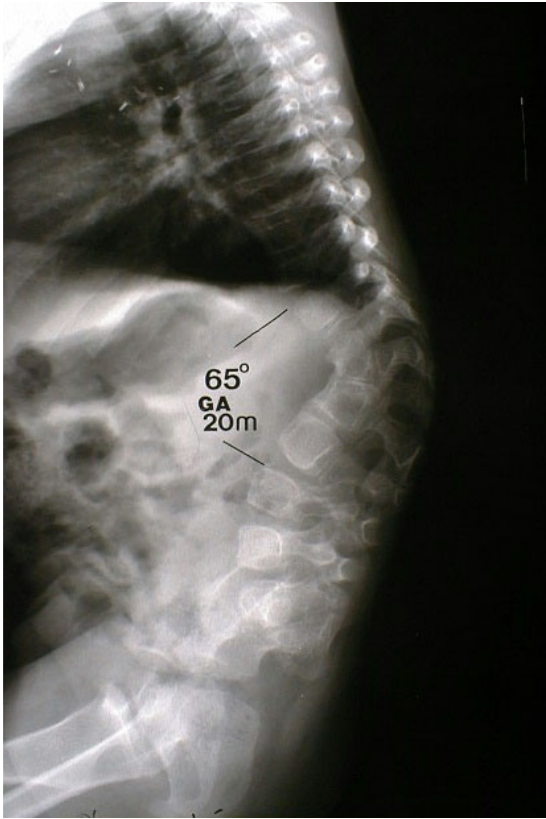


SEVERE

# FULL ANTERIOR FUSION

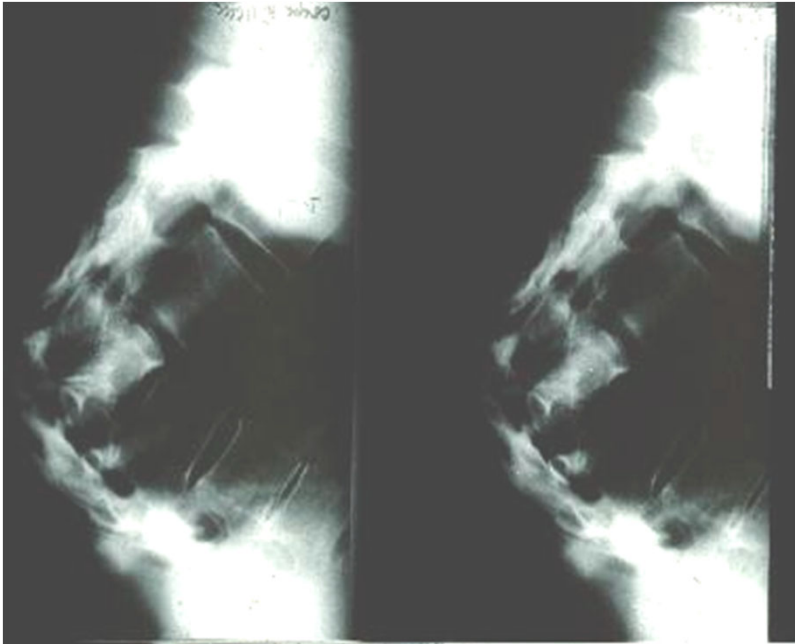


# FULL ANTERIOR FUSION



## FULL ANTERIOR FUSION

35 Years of age  
Paraplegia after a jump



Mechanical and vascular  
injury

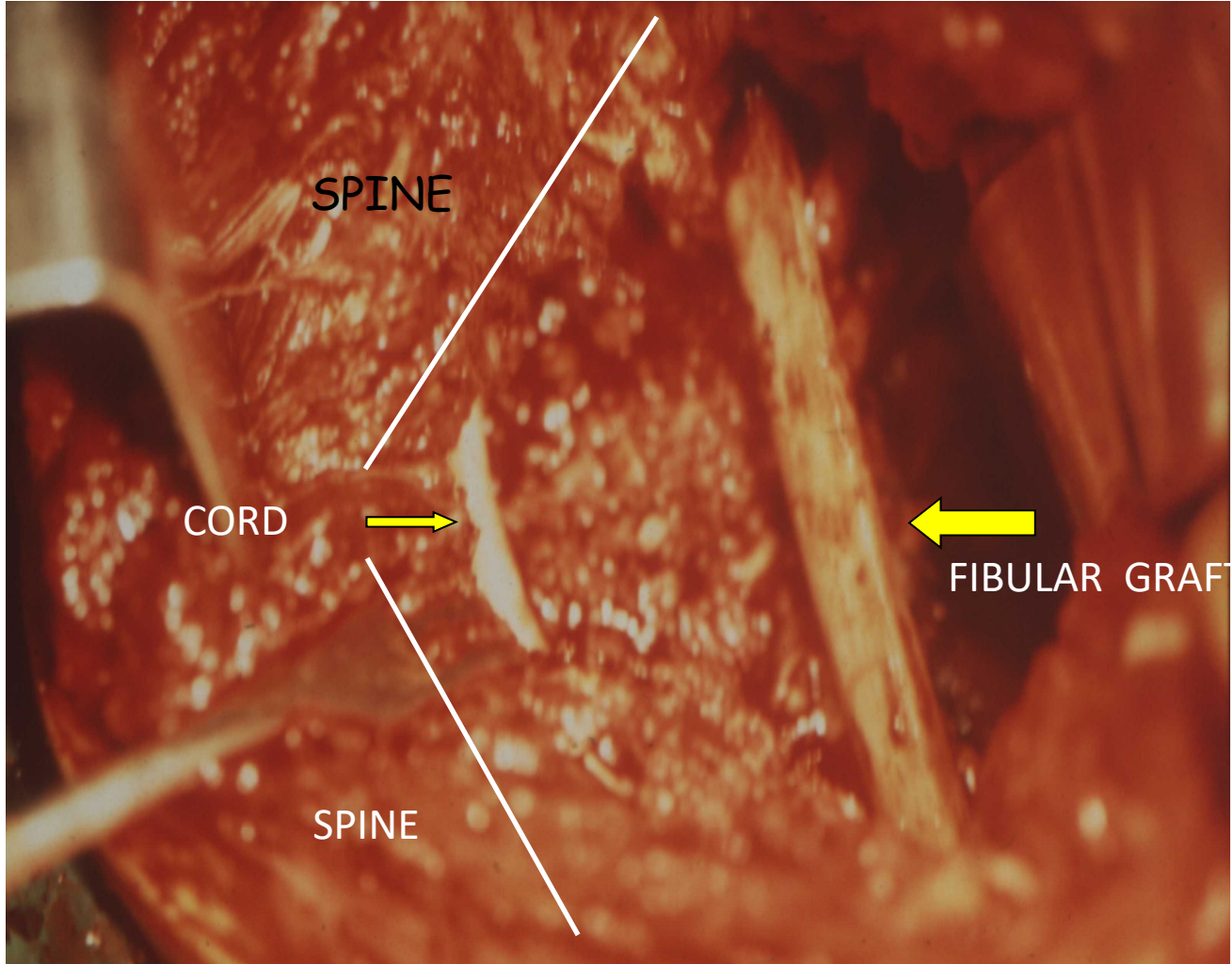
Courtesy J Dubousset

## FULL ANTERIOR FUSION



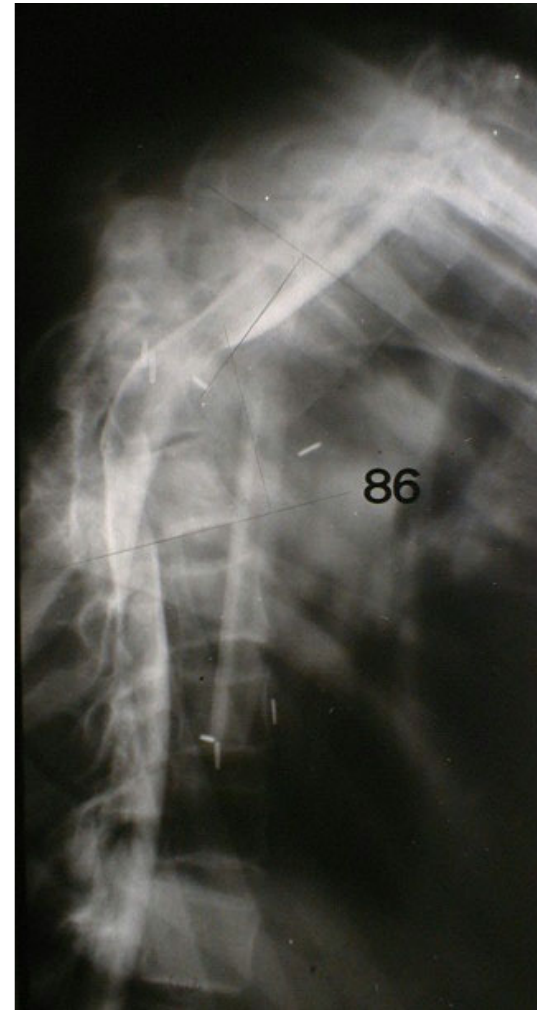
Progressive non traumatic  
spastic paraplegia

**FULL ANTERIOR FUSION**



INTRA THORACIC CORD TRANSLOCATION

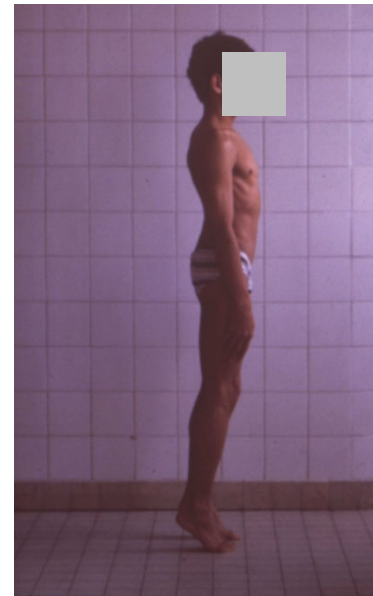
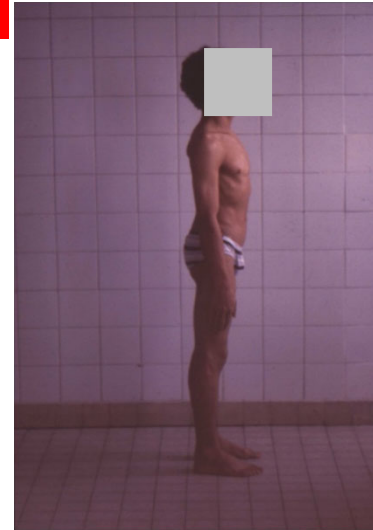
**FULL ANTERIOR FUSION**



INTRA THORACIC CORD TRANSLOCATION

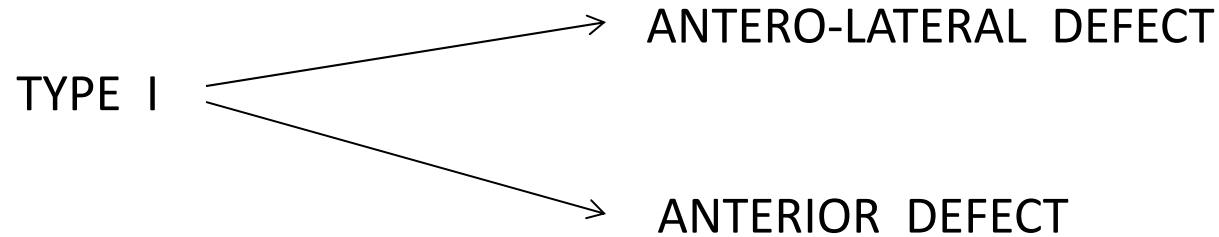


# FULL ANTERIOR FUSION



## MODIFIED WINTER CLASSIFICATION

### DEFECT of FORMATION



### DEFECT of SEGMENTATION



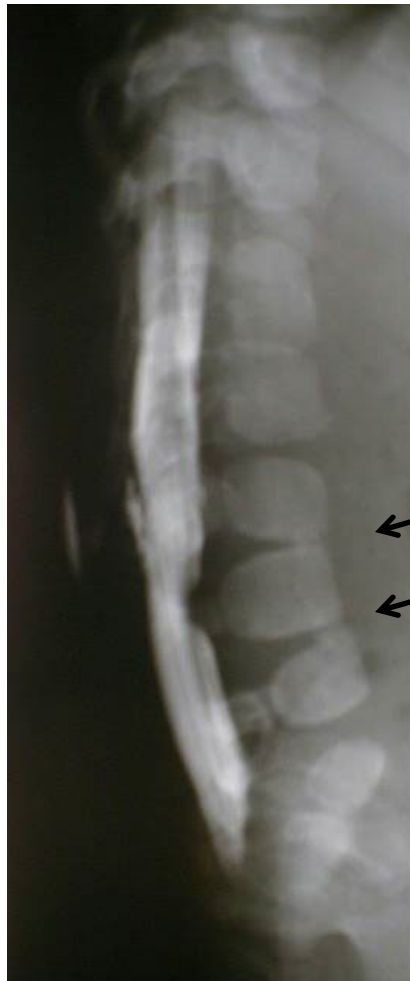
### COMPLEXE MALFORMATION

TYPE III

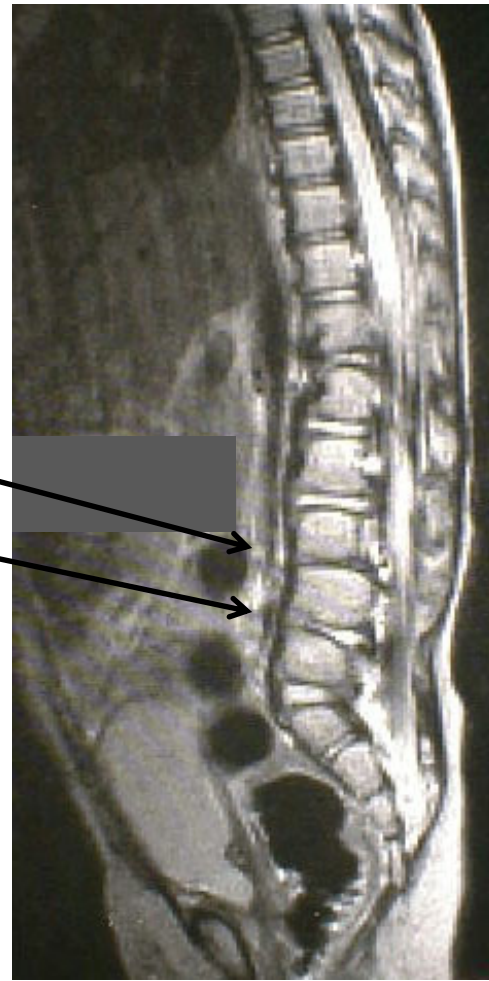
# PROGRESSIVE ANTERIOR FUSION

Progressively ossifying anterior unsegmented bar

1 Y



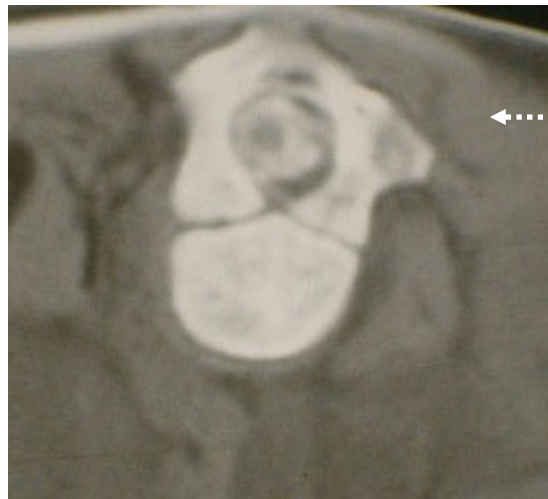
Disc spaces  
L 3 L 4  
L 4 L 5  
Start to ossify anteriorly



## PROGRESSIVE ANTERIOR FUSION

Associated spinal malformations

Narrowing canal at the level of the bar



# PROGRESSIVE ANTERIOR FUSION

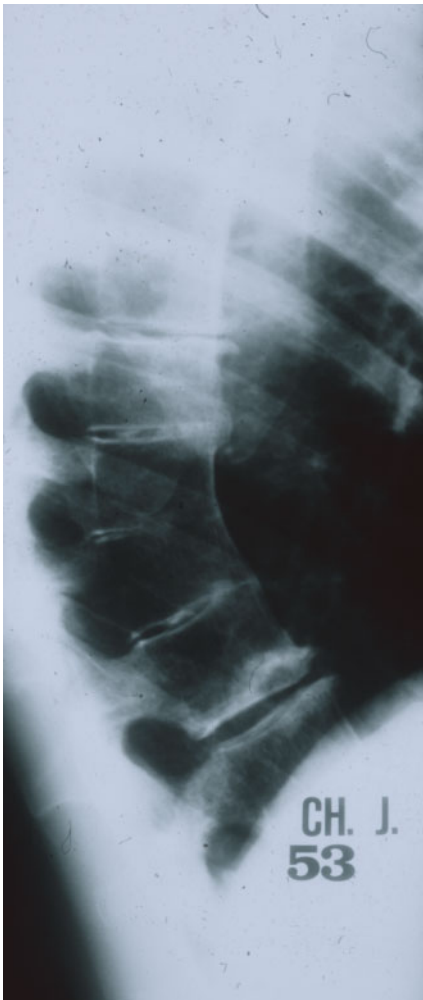
## NATURAL HISTORY

1 Y



15 Y

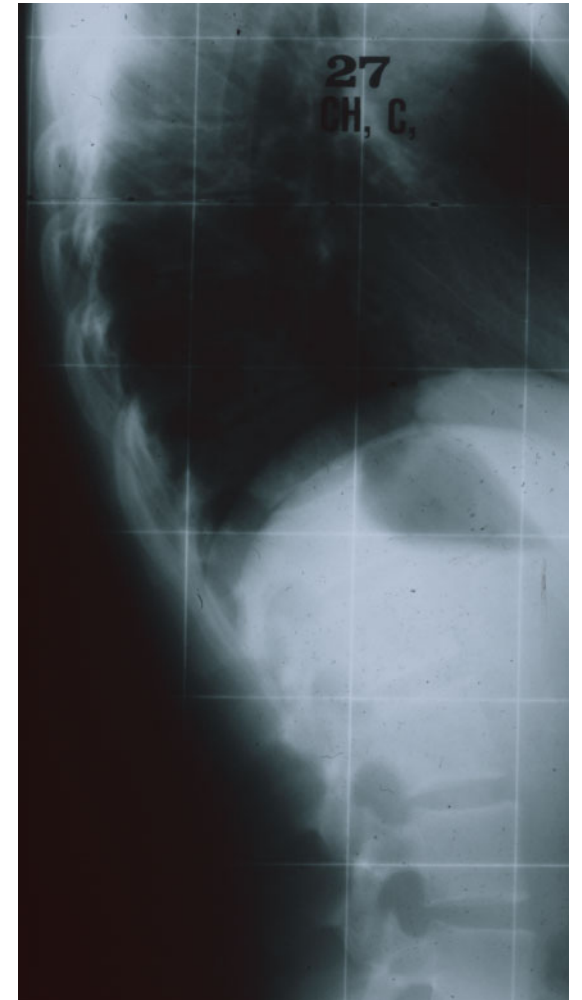




Jacqueline 53 Y  
The mother



Sophie 29 Y  
First daughter



Catherine 27 Y  
Second daughter

## PROGRESSIVE ANTERIOR FUSION

Case	At Presentation			Treatment Free		Rate of Annual Progression
	Age	Bar's Location	Segmental Angle	Segmental Angle at Follow-up	Follow-up	
1	2Y 3M	L: T12-S1	41°	44°	1Y 2M	2.6°
2	7Y 8M	T: T7-T12	47°	53°	2Y 6M	2.4°
3	5Y 3M	T: T9-T10	14°	22°	20Y 7M	0.4°
4	8Y	L: T11-L4	85°	85°	0	–
5	14Y	TL: T10-L1	40°	40°	3Y	0°
6	3Y	L: L1-L5	33°	92°	12Y	4.9°
7	14Y 8M	L: L1-L4	72°	72°	2Y 7M	0°
8	16Y 8M	T: T9-T11	40°	40°	0	–
9	11Y 4M	T L: T10-T12	35°	30°	12Y 2M	-0.4°
10	10Y 5M	L: L1-L3	14°	13°	2Y 10M	-0.4°
11	13Y 3M	TL: T10-T12	17°	17°	0	–
12	13Y 4M	TL: T9-T12	22°	23°	3Y 11M	0.3°
13	4Y 11M	L: L3-L5	- 20°	6°	5Y 11M	4.4°
14	1Y	L: L3-L5	8°	8°	11M	0°
15	1Y 1M	L: L1-L3	16°	14°	1Y 1M	-1.8°
16	3M	L: L1-L3	20°	18°	1Y 2M	-1.7°

### Natural History of Segmental Angle Progression

**Progressive Anterior Vertebral Bars: A Study of 16 Cases.**

Bollini, Gerard; MD, PhD; Guillaume, Jean-Marc; Launay, Franck; Zeller, Reinhard; Jouve, Jean-Luc; MD, PhD; Viehweger, Elke; Katchburian, Marcos; Dubousset, Jean; MD, PhD  
Spine. 36(6):E423-E428, March 15, 2011.

## PROGRESSIVE ANTERIOR FUSION

Case	At Presentation			Treatment Free		Rate of Annual Progression
	Age	Bar's Location	Segmental Angle	Segmental Angle at Follow-up	Follow-up	
3	2Y 3M	T: T9-T10	14°	22°	20Y 7M	0.4°

Thoracic T9 T 10 at 2 Y 3 M

20 Years Follow up

Kyphosis angle increase from Fourteen to Twenty two degrees

### Natural History of Segmental Angle Progression

**Progressive Anterior Vertebral Bars: A Study of 16 Cases.**

Bollini, Gerard; MD, PhD; Guillaume, Jean-Marc; Launay, Franck; Zeller, Reinhard; Jouve, Jean-Luc; MD, PhD; Viehweger, Elke; Katchburian, Marcos; Dubousset, Jean; MD, PhD  
Spine. 36(6):E423-E428, March 15, 2011.



## PROGRESSIVE ANTERIOR FUSION

Case	At Presentation			Treatment Free		Rate of Annual Progression
	Age	Bar's Location	Segmental Angle	Segmental Angle at Follow-up	Follow-up	

6	3Y	L: L1-L5	33°	92°	12Y	4.9°
---	----	----------	-----	-----	-----	------

Lumbar L 1 L 5 at 3 Y

12 Years Follow up

Kyphosis angle increase from Thirty Three to Ninety two degrees

### Natural History of Segmental Angle Progression

**Progressive Anterior Vertebral Bars: A Study of 16 Cases.**

Bollini, Gerard; MD, PhD; Guillaume, Jean-Marc; Launay, Franck; Zeller, Reinhard; Jouve, Jean-Luc; MD, PhD; Viehweger, Elke; Katchburian, Marcos; Dubousset, Jean; MD, PhD  
Spine. 36(6):E423-E428, March 15, 2011.

## PROGRESSIVE ANTERIOR FUSION

### NATURAL HISTORY

- The evolution depends upon the location of the bar and the age of the patient
- In the thoracic and the thoraco-lumbar area , low risk for progression
- In the lumbar area the risk for progression is very high

# PROGRESSIVE ANTERIOR FUSION

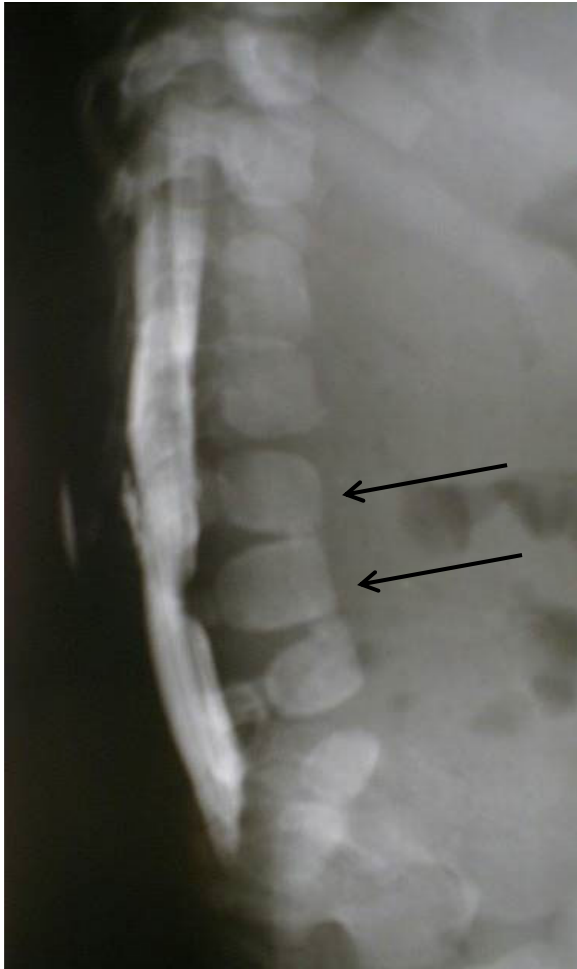
## NATURAL HISTORY



LEADS TO MODERATE ANGULATION  
AT THE THORACIC LEVEL

## PROGRESSIVE ANTERIOR FUSION

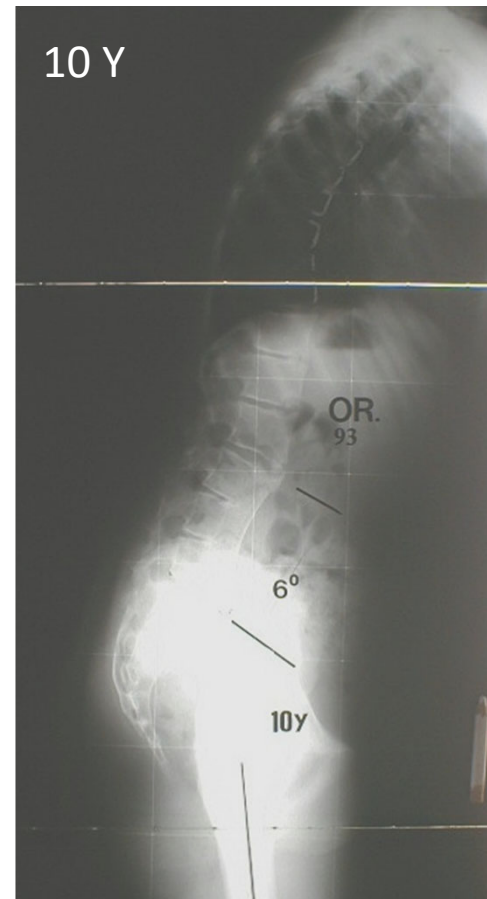
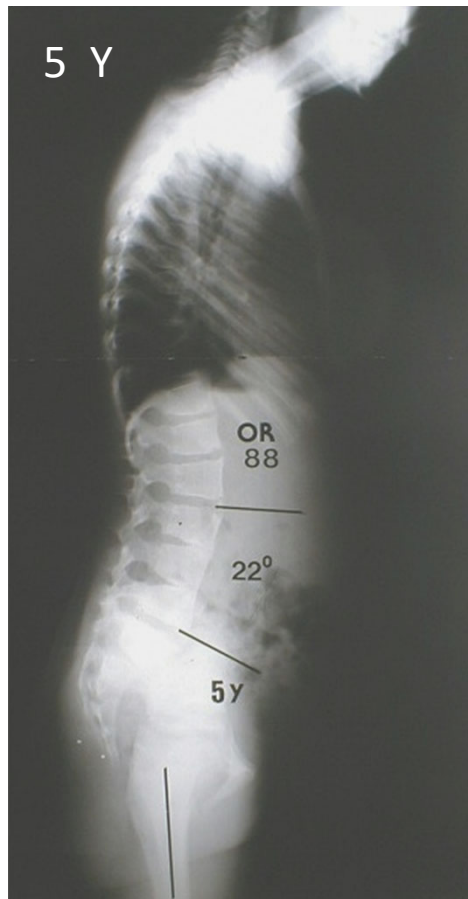
### NATURAL HISTORY



LEADS TO MAJOR ANGULATION  
AT THE LUMBAR LEVEL

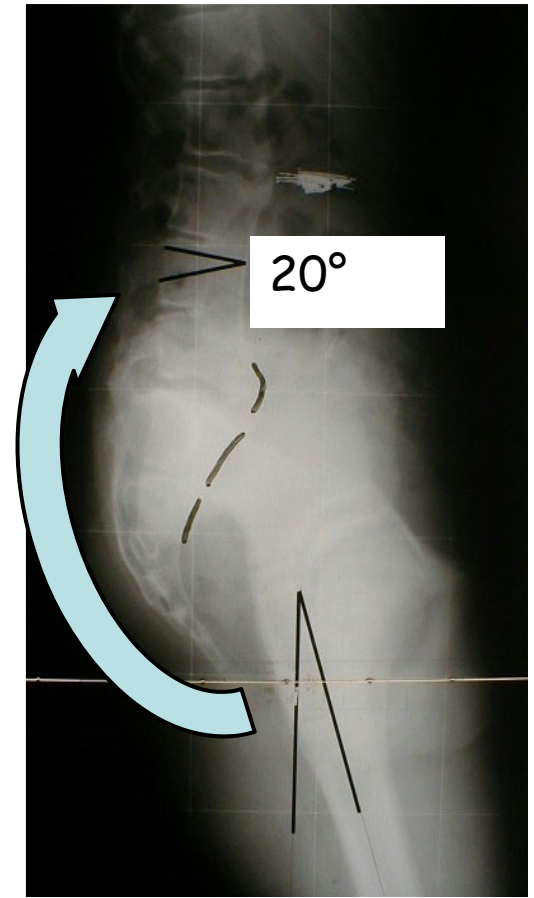
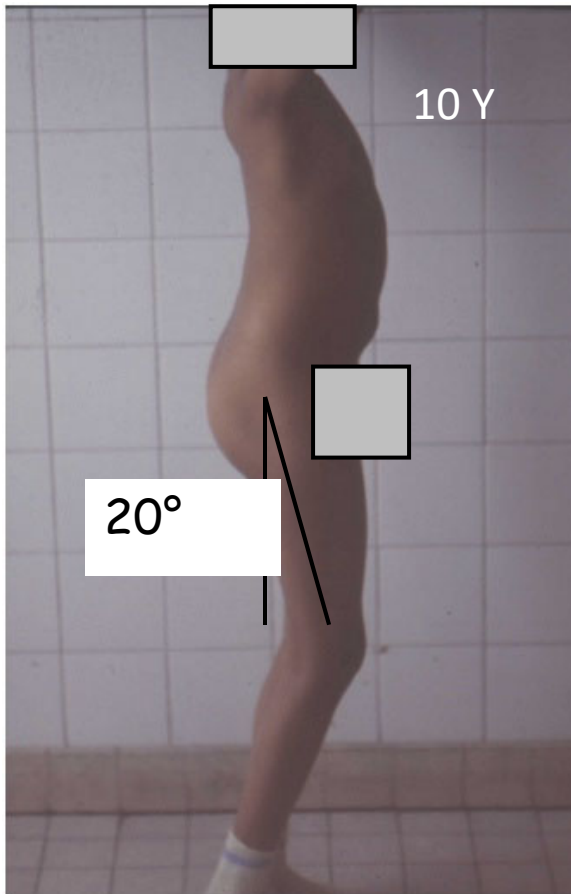
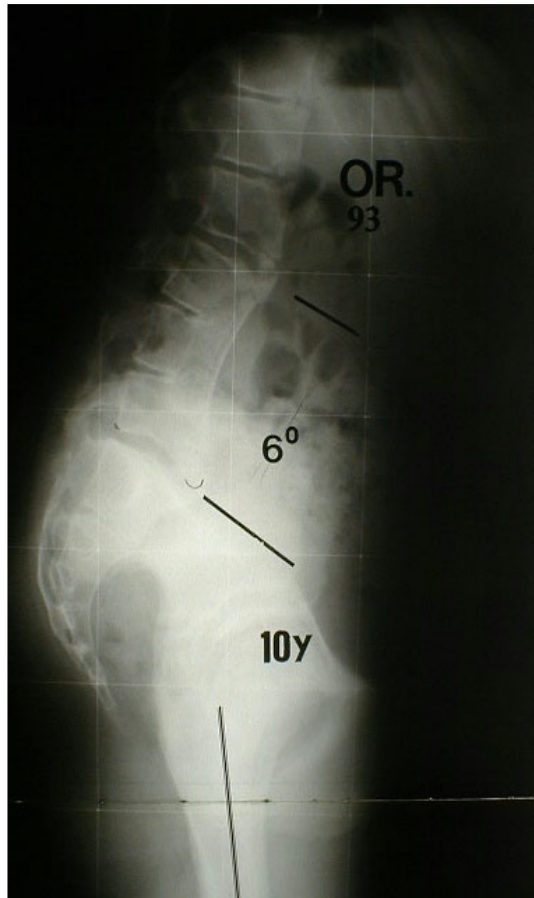
# PROGRESSIVE ANTERIOR FUSION

## CORRECTION OF LATE DEFORMITIES



# PROGRESSIVE ANTERIOR FUSION

## CORRECTION OF LATE DEFORMITIES



# PROGRESSIVE ANTERIOR FUSION

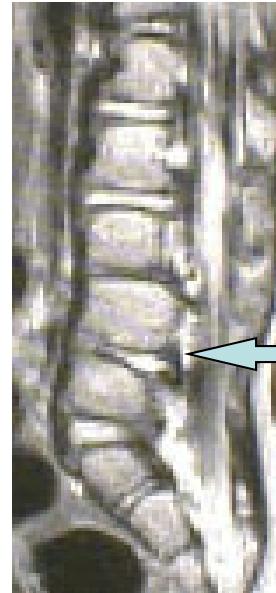
## CORRECTION OF LATE DEFORMITIES



## PROGRESSIVE ANTERIOR FUSION

### Hypothesis

- Progressive ossification runs from anterior to posterior destroying a normal disc structure
- Removal of the bar with interpositional material would lead to a normal further growth of the remaining disc structure

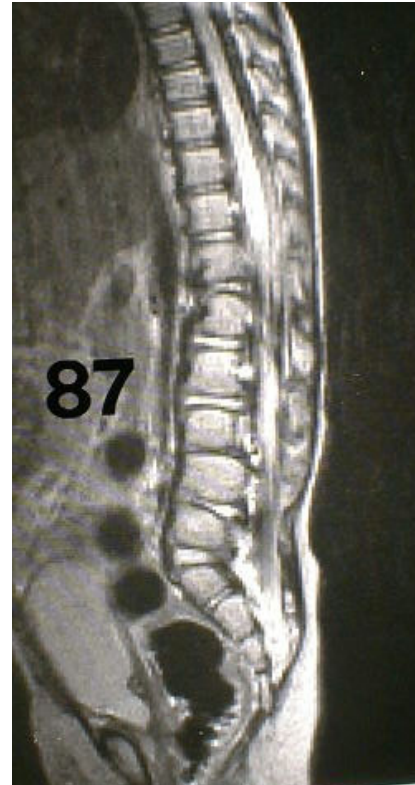
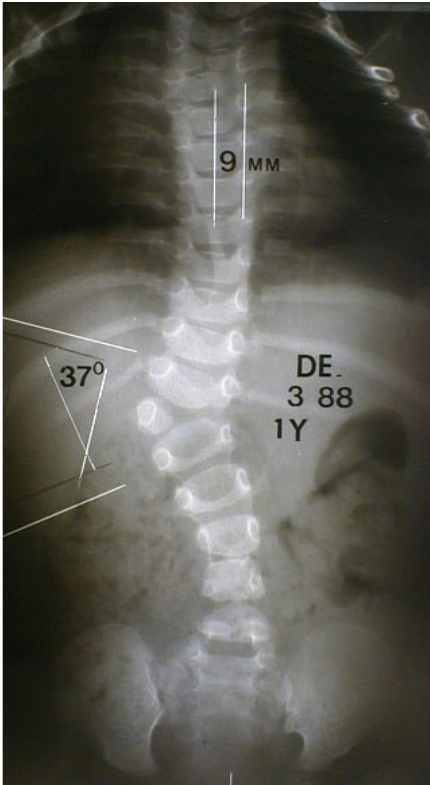


Normal T2  
Weighted  
MRI Signal



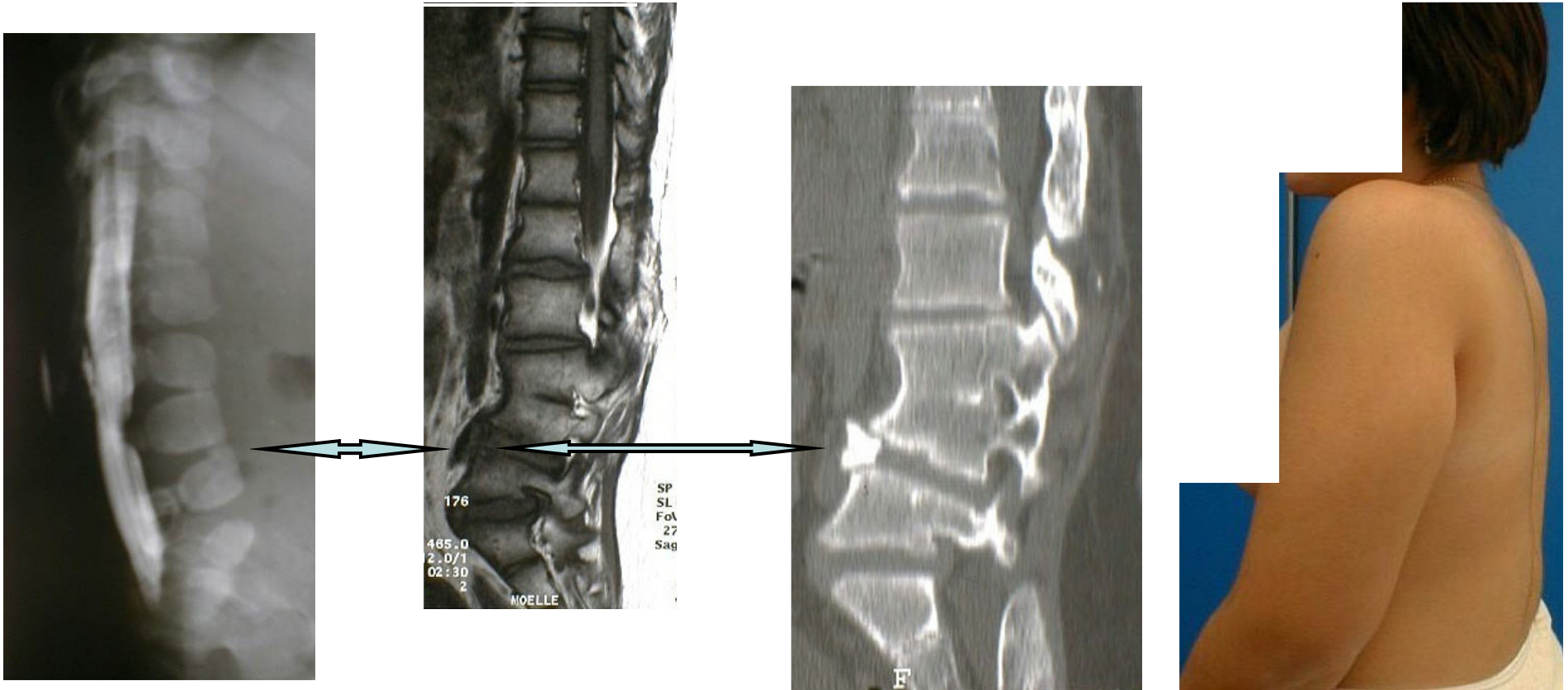
# PROGRESSIVE ANTERIOR FUSION

## PREVENTION OF LATE DEFORMITIES



# PROGRESSIVE ANTERIOR FUSION

## PREVENTION OF LATE DEFORMITIES



# PROGRESSIVE ANTERIOR FUSION

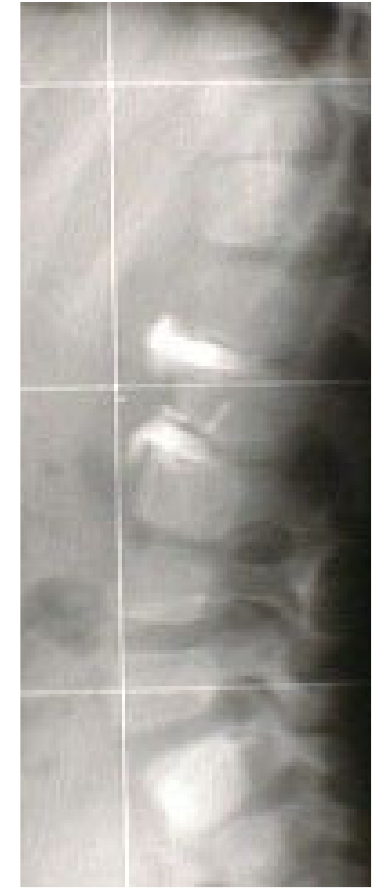
## PREVENTION OF LATE DEFORMITIES



PRE-OP



J+2ANS



J+4ANS

## PROGRESSIVE ANTERIOR FUSION

### MATERIAL

- 5 patients, 3 Females, 2 Males
- Mean age at diagnosis 21 months [12-60]
- Associated vertebral malformation in the whole series

**PROGRESSIVE ANTERIOR FUSION****MATERIAL**

Patient	Sexe	Age at diagnosis	Location	Discs number involved
Via. A	F	5 Y	L4-L5	1
Ver. T	M	16 M	T12-L1	1
Dem. J	F	12 M	L3-L5	2
Ger. A	F	13 M	L1-L3	2
Pey. A	M	8 M	L1-L3	2

**PROGRESSIVE ANTERIOR FUSION****MATERIAL**

Patient	Sexe	Age at diagnosis	Age at surgery	F.U.
Via. A	F	5A	5 A 8 M	3 A
Ver. T	M	16M	2 A 7 M	2 A 8 M
Dem. J	F	12M	1 A 11 M	17 A
Ger. A	F	13M	2 A 2 M	11 A
Pey. A	M	8M	1 A 7 M	7 A 9 M

## PROGRESSIVE ANTERIOR FUSION

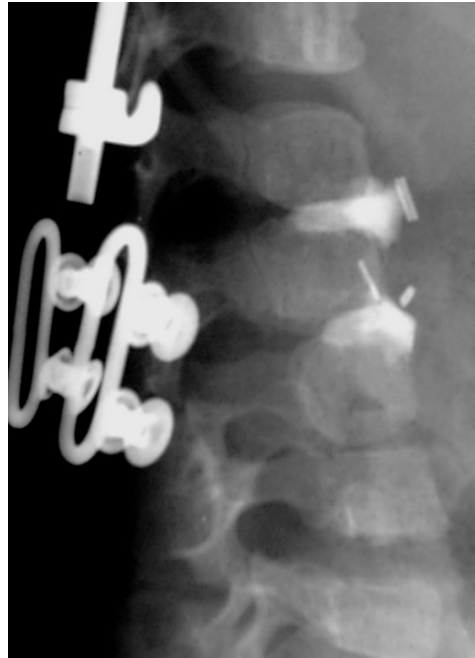
### RESULTS

- Mean age at surgery 2 Y 2 M (1 Y 7 M- 5 Y 8 M)
- Mean F.U. 8 Y 1 M (2 Y 8 M-17 Y)
- Pre op. mean kyphotic segmental angle: 18,5° (12-34°)
- Post op. mean kyphotic segmental angle: 14,5° (6-28°)
- Mean segmental kyphotic angle at F.U.: 12° (0°-26°)

**PROGRESSIVE ANTERIOR FUSION**



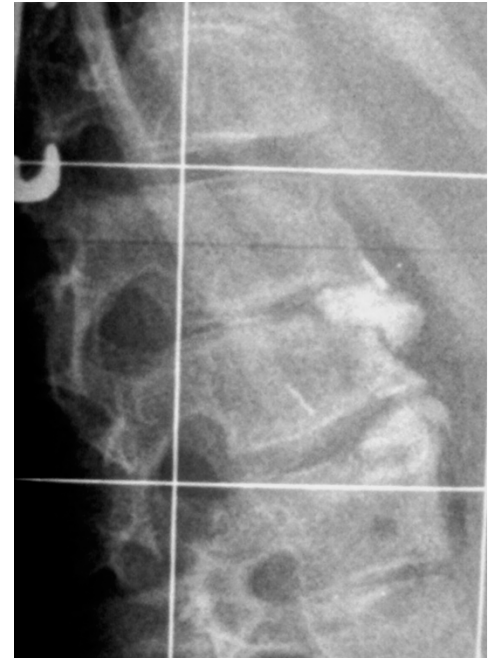
4 M



1 A 7 M



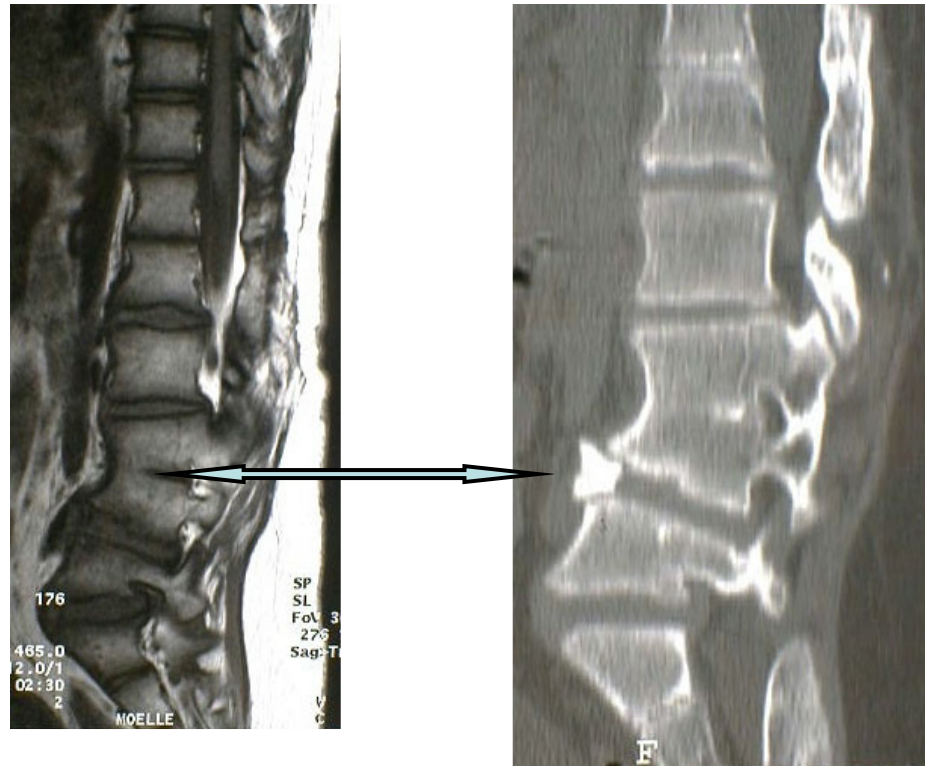
3 A 1 M



8 A



# PROGRESSIVE ANTERIOR FUSION



MARSEILLE

THANK YOU

