Neuromuscular Scoliosis

Mike Gibson

Newcastle, Feb 2009

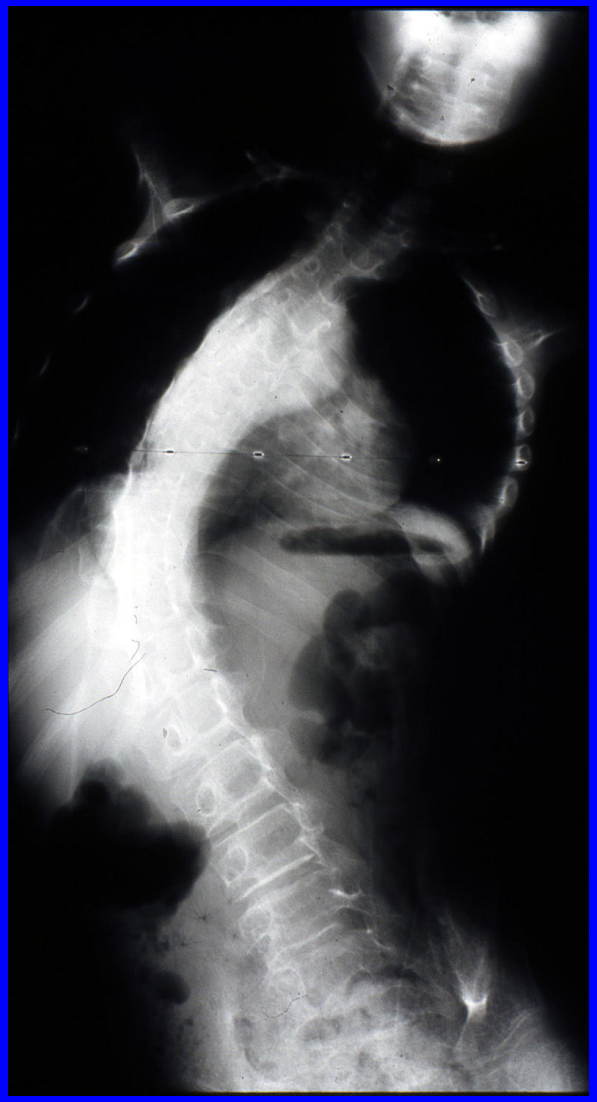
Neuromuscular Scoliosis

The aim of treatment is to maximise Function.

Neuromuscular Scoliosis

Characteristic features;

- Progressive
- Long involving most of the thoraco-lumbar spine
- Initially flexible rapidly become stiff
- Unbalanced



Problems in the treatment of Neuromuscular scoliosis;

- Severe soft tissue contractures, poor bone.
- Unfit patients with associated medical problems (respiratory & cardiac).
- Uncooperative patients.
- Neurological – epilepsy or involuntary movements.
- Poor nutritional status

Classification of neuromuscular scoliosis- Neurological

Upper motor neurone

- Cerebral Palsy
- Spinocerebellar degeneration-Fredericks ataxia, Charcot Marie Tooth, Roussy-Levy.
- Syringomyelia
- Spinal cord tumour
- Spinal cord trauma

Lower motor neurone

- Poliomyelitis
- Other viral myelitides
- Traumatic
- Spinal muscular atrophy-Werdig-Hoffmann , Kugelberg-Wellander
- Dysautonomic disorders (Riley-Day syndrome)

Classification of neuromuscular scoliosis- Muscular

- Arthrogryposis
- Muscular Dystrophy- Duchenne, Limb-Girdle, Facio-scapulo-humeral
- Fibre- type disproportion
- Congenital hypotonia
- Myotonia dystrophica

Assessment of patients with neuromuscular scoliosis

Clinical Assessment

- Level of function- ambulation, sitting balance, head control, intellectual ability
- Spine- sitting & prone, curve flexibility, pelvic obliquity.
- Examine hips.
- Neurological status

Assessment of Lung Function

- History of chest infections
- Examination of chest (Cough reflex)
- Chest X-ray
- Lung Function Tests (Compliant patients)- Spirometry, Sniff nasal inspiratory pressures
- Overnight oxygen saturation
- Sleep studies

SNIFF NASAL INSPIRATORY PRESSURE [SNIP]

- non-invasive test
- nasal pressure measured in occluded nostril during a maximal sniff through the contralateral nostril.
- Sniff is a physiological manoeuvre, easily performed and reproducible in children.
- SNIP is the most accurate & reproducible test to assess global inspiratory muscle strength, especially diaphragmatic strength.

Polkey 1995

Assessment of Cardiac and Nutritional status

Cardiac

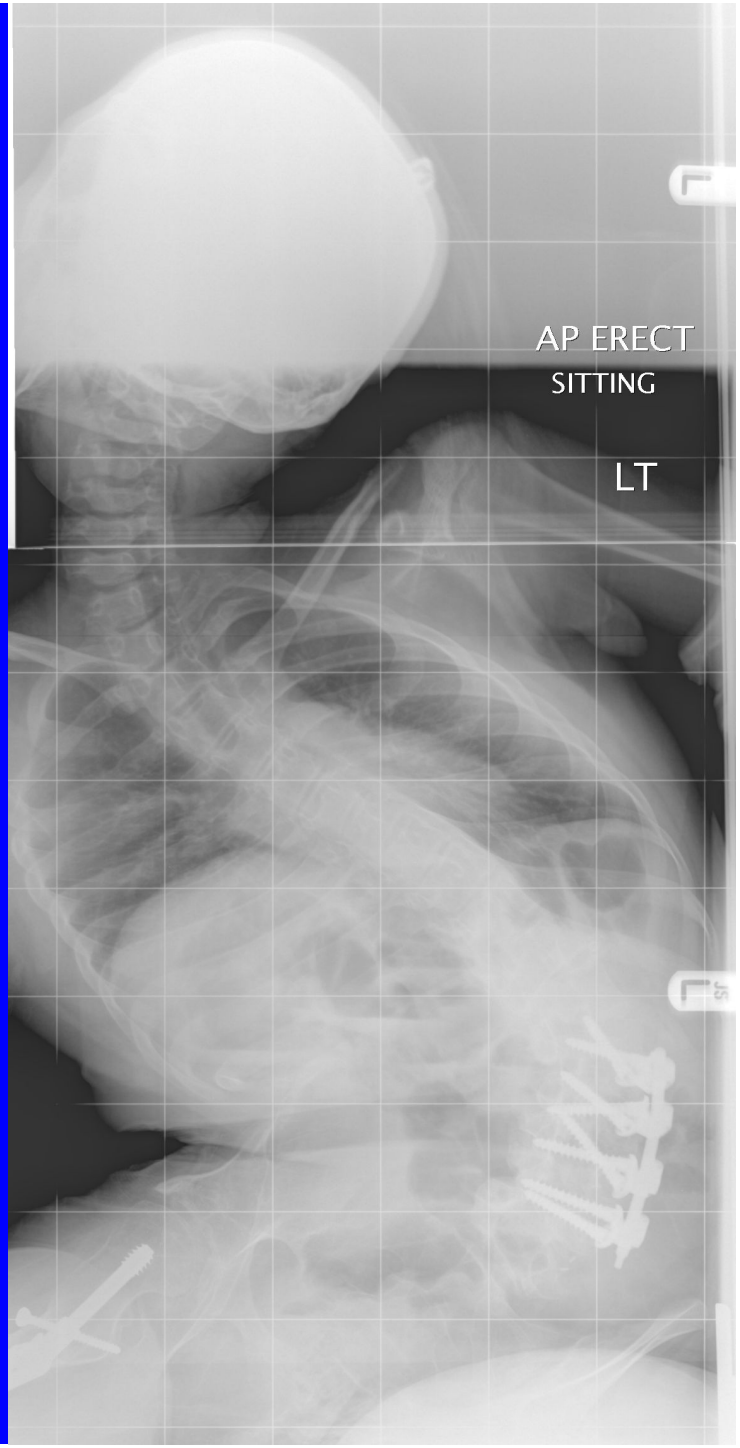
- CXR
- ECG
- Echocardiogram

Nutritional

- Total protein
- Albumin (3.5 g/l)
- Lymphocyte count

Imaging of the Spine

- Erect radiographs (Standing/sitting)- AP and Lateral views.
- Flexibility tests- Traction views, prone push pull views.
- Additional imaging with CT or MRI rarely required.
- BMD



Selection of Treatment Option

- Level of function.
- Understanding of natural history of underlying condition.
- Life expectancy.
- Intellectual function. Important in helping the family decide on treatment option.

Treatment Options in Neuromuscular Scoliosis

Non operative treatment

- Wheelchair modification
- Bracing- Underarm braces hard or soft

Surgery

- Posterior instrumentation and fusion

Bracing in Neuromuscular Scoliosis

- No evidence bracing alters natural history.
- Braces may delay progression.
- Braces improve sitting balance in Flexible curves

Surgery in neuromuscular scoliosis

Objectives-

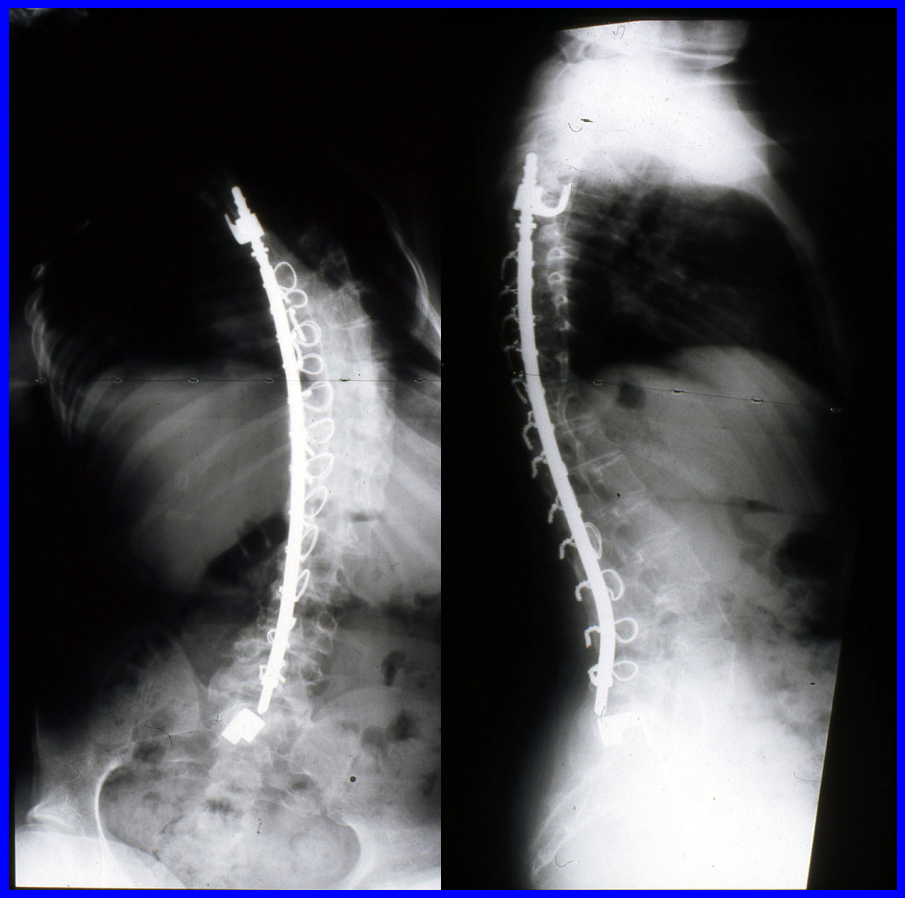
- A balanced spine in the frontal and sagittal planes;
- A level pelvis;
- A solid fusion.

Principles

- Instrument and fuse long –risk of junctional deformities.
- End fusion at L5 or sacrum

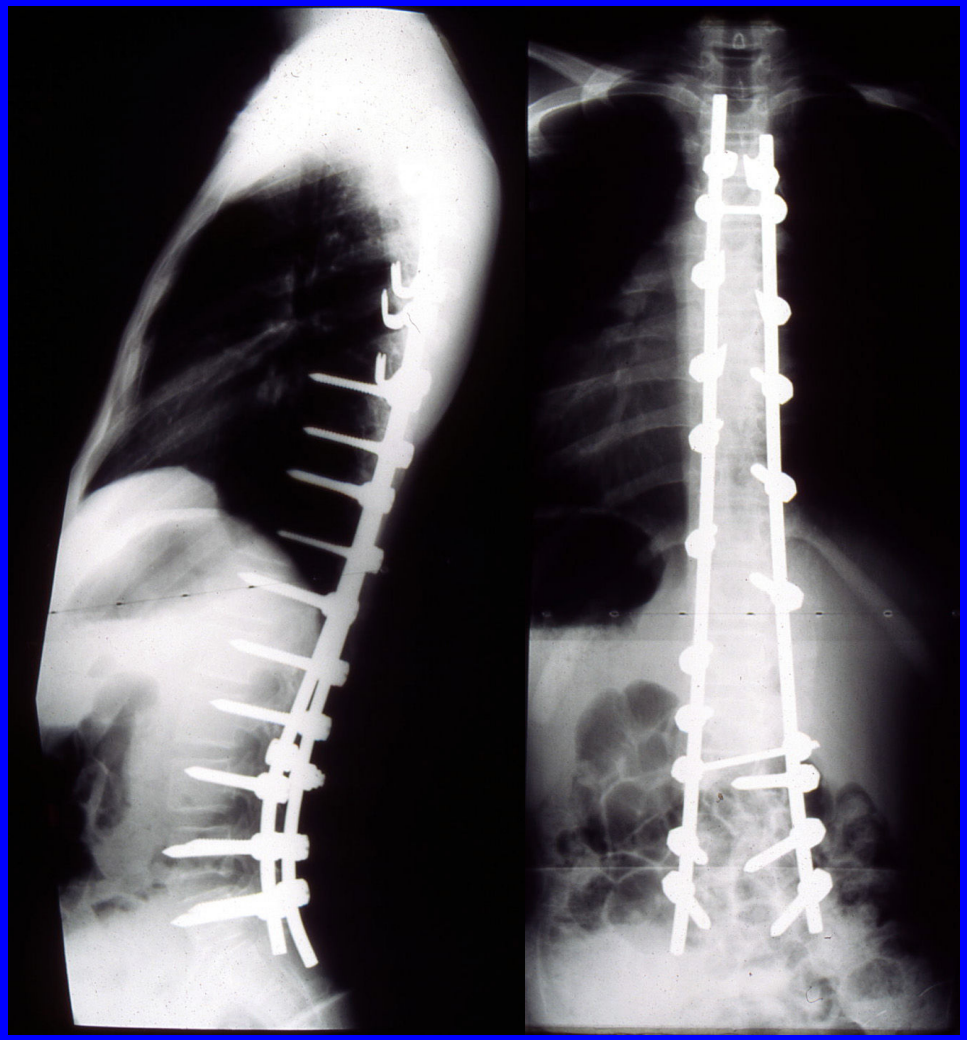
Neuromuscular Scoliosis Choice of Instrumentation

- Harrington rods
- Harrie- Luque
- Luque Galveston
- Universal systems pedicle screws / hooks



Neuromuscular Scoliosis Choice of Instrumentation

- Fast
- Simple
- Rigid
- Safe
- Cheap



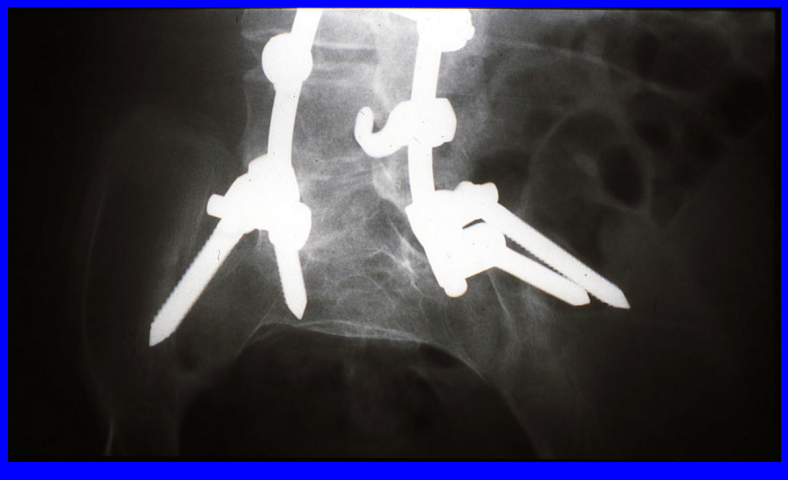
Selection of distal fixation level

To L5;

- Ambulant patients.
- Flexible curves.
- <10 degrees pelvic obliquity on traction films.

To sacrum;

- Non ambulant patients.
- >10 degrees fixed pelvic obliquity



Surgical technique neuromuscular scoliosis

Allograft bone-

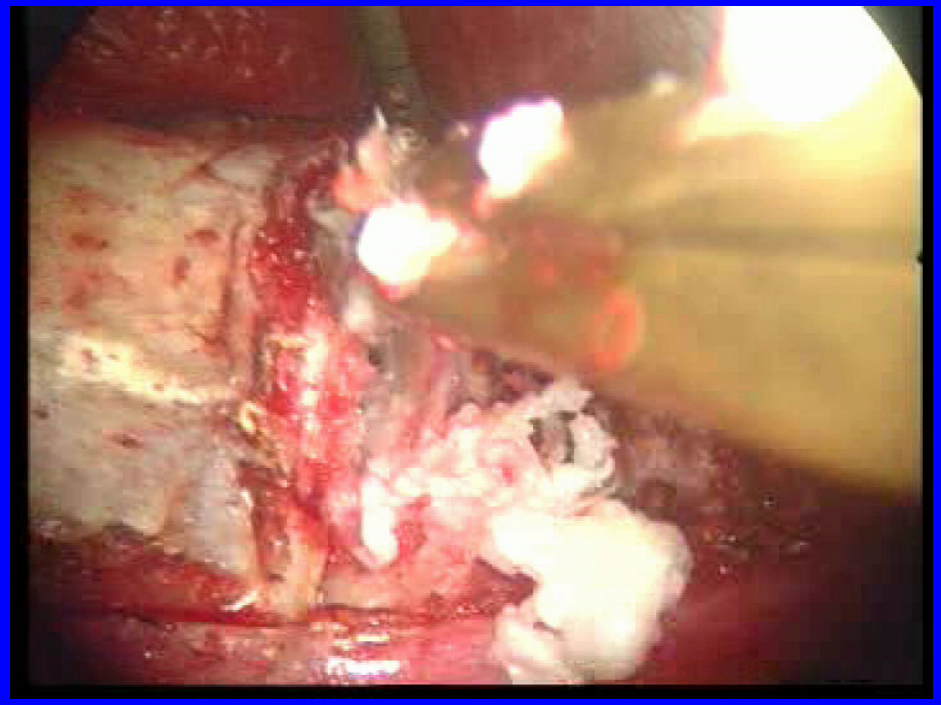
- large amount of graft needed,
- Posterior Pelvis may be needed to anchor fixation,
- speed.

X matched blood, cell saver

SSEP monitoring

Anterior surgery in neuromuscular scoliosis

- Anterior release in severe stiff curves.
- To prevent crankshaft
- Limited role for anterior instrumentation.



Scoliosis in Duchenne Muscular Dystrophy

Aetiology of DMD;

- X- Linked recessive Genetic disease.

Aetiology of scoliosis in DMD;

- Muscle imbalance and weakness developing before skeletal maturity.

Scoliosis in Duchenne Muscular Dystrophy;

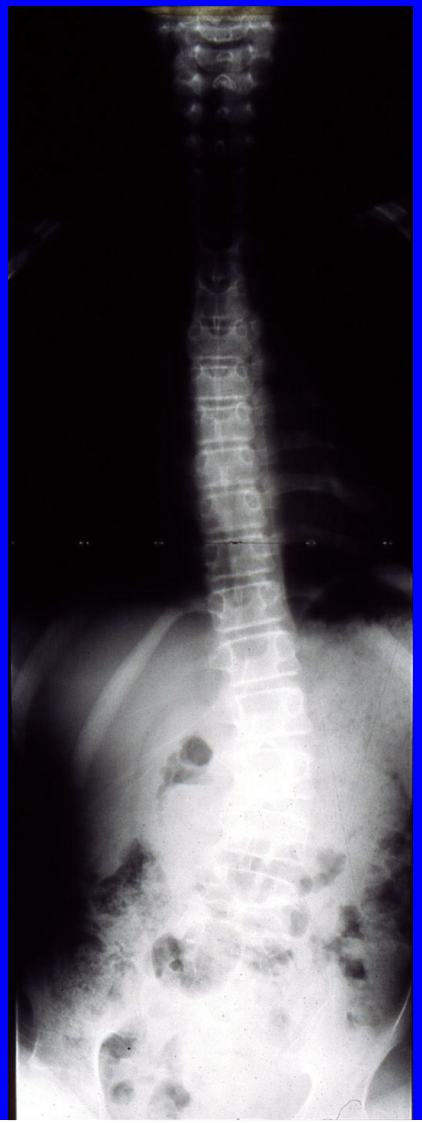
- 95% develop Scoliosis
- Develops 1-2 yrs after going “Off feet”
- Progressive
- Affects sitting balance
- Reduces pulmonary function
- Decreases quality of life.

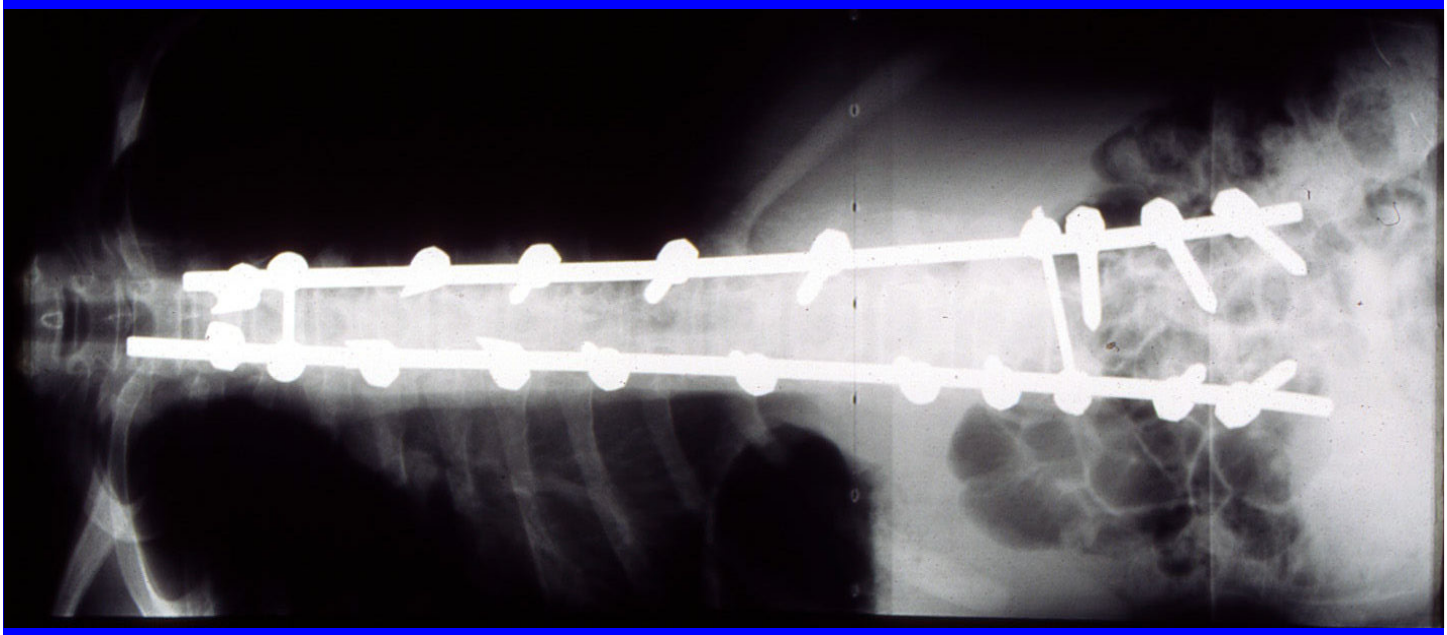
Surgery in DMD

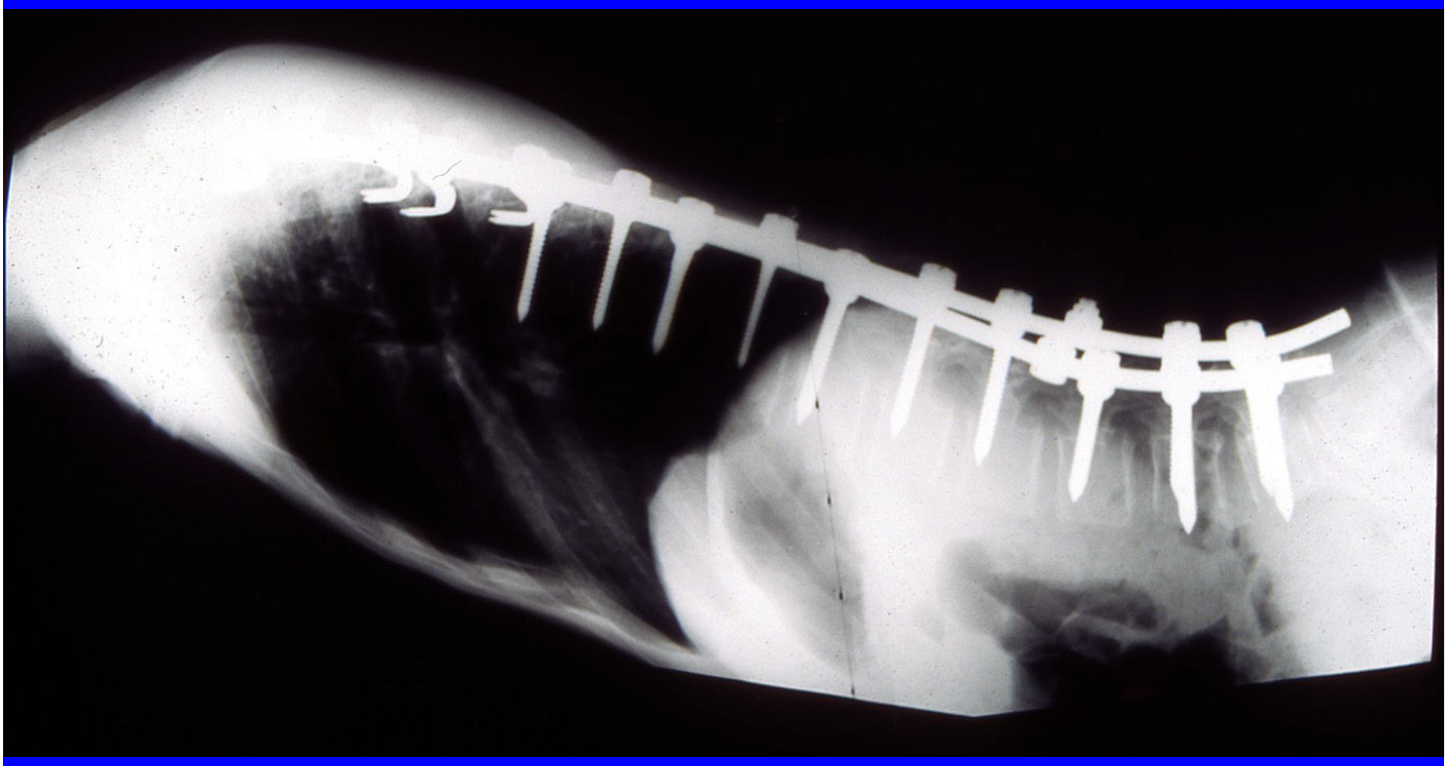
- Technically demanding surgery;
- Initially surgeons reluctant to operate;
- Late surgery on severe scolioses;
- High complication rate in early series;
- 8 L av. blood loss

Surgery in Duchenne M.D.

- Operate early.
- 20 degree curves.
- Fusion to L5.
- Less blood loss.
- Better correction.
- Improved outcome







Lung function in DMD: Changes in FVC

↓ 4 % / year

↓ 4 % / 10° **Kurz, JPO 1983**

> 35°, ↓ 40 % predicted

Smith, JBJS 1989

Respiratory failure:
Pathogenesis

- ↓ Chest wall compliance,
- Micro-atelectasis,
- Nocturnal hypoventilation.

Nocturnal hypoventilation:
Clinical effects

- Frequent waking at night,
- Un-refreshing sleep
- Daytime somnolence

- **Morning headaches and nausea (hypercapnia)**

Home ventilation team

- FVC ↓ 1 litre,

- Chest infections,
- Sleep pattern,
- Appetite & weight loss.

- Overnight oximetry.


BREAS

Objective

To assess the combined effect of surgery and nocturnal ventilation on lung function and survival patterns in patients with DMD.

91 patients with DMD:

1986 - 2002

39 Spinal fusion

52 No surgery

52 patients: No spinal surgery

- Reluctant surgeon,
- Refused by patient / family,
- Refused by anaesthetist: cardiomyopathy,
- No scoliosis.

39 patient: Spinal fusion

- **Loss of ambulation: 9.6y**
- **Referred at: 12.5y**
- **Age at operation: 14.5y**

Spinal fusion technique

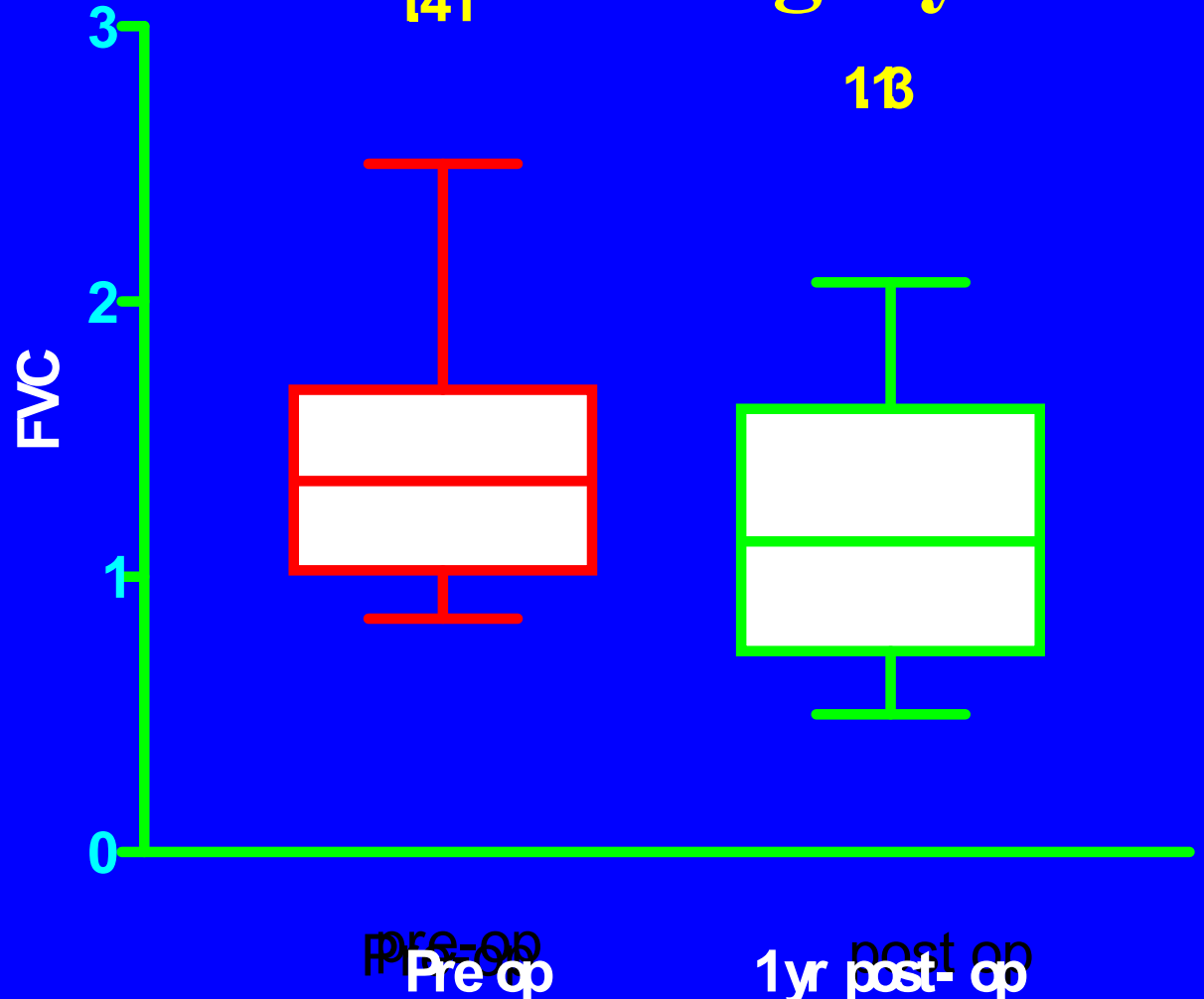
- 2 surgeons
- 32 AO USS
- 3 Cotrel-Dubosset
- 3 Colorado
- 1 Harrie- Luque

Respiratory Function;

Initial reduction in pulmonary function,which then returns to pre op. level and then gradually decreases.

Kennedy et al (1995), Miller et al (1991)

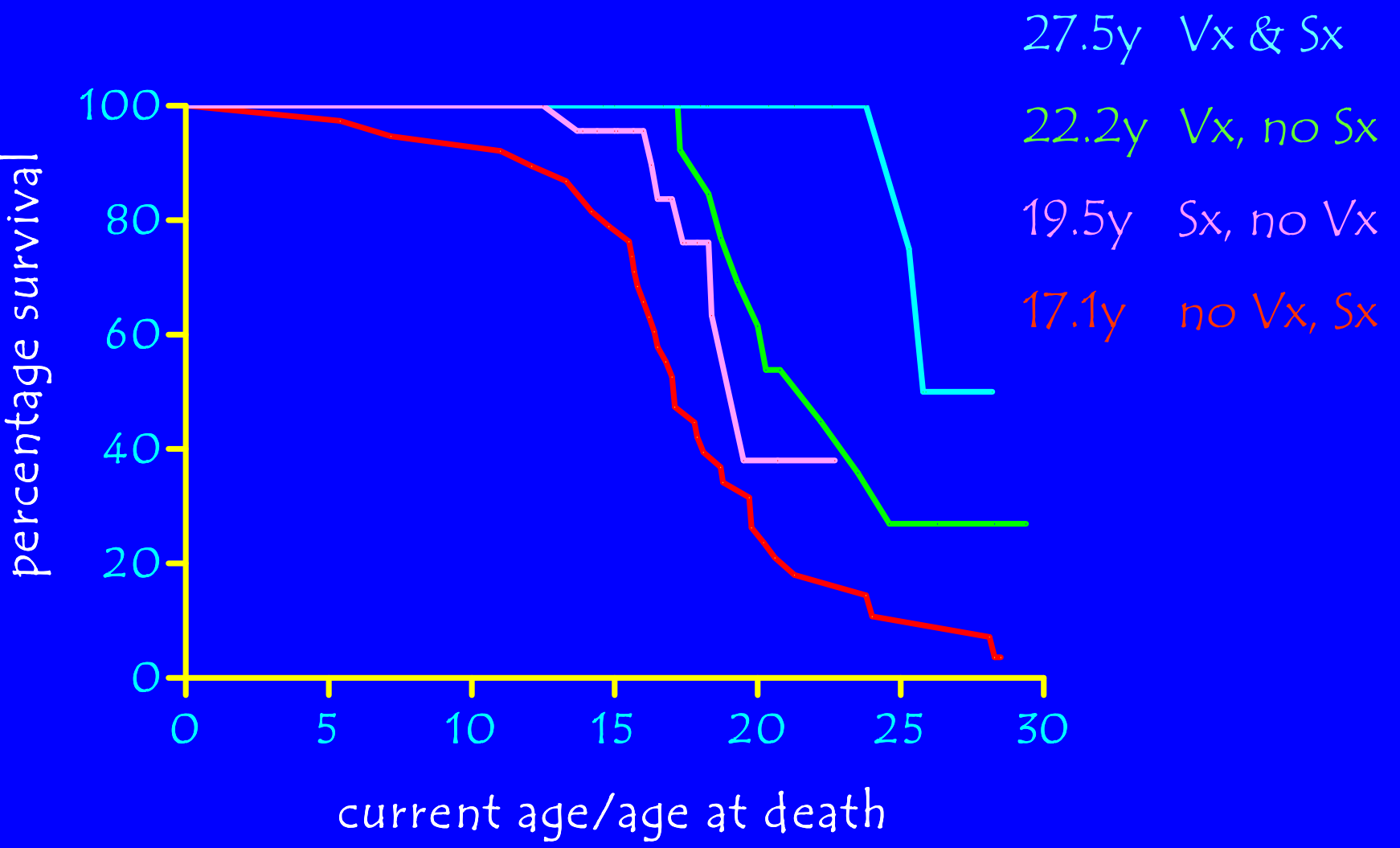
Effect of surgery on FVC



- Changing surgical practice, over the 10 years, gradually becoming more surgically aggressive
- Gradual introduction of ventilation,
- 4 groups to assess survival.

- Spinal surgery and nocturnal ventilation, 15
- Nocturnal ventilation (no spinal surgery), 14
- Spinal surgery (no ventilation), 24
- Neither spinal surgery nor ventilation, 38

Kaplan Meier analysis



Cardiomyopathy

- Median survival 16.3y,
- Main anaesthetic contra-indication.

Conclusion

- Longest survival: ventilation and surgery,
- Ventilation alone better than surgery alone,
- Worst survival: Early symptomatic cardiomyopathy.

Scoliosis in DMD

- The Zenith for surgery!

- Steroid therapy.
- Gene therapy.

Steroids in DMD

Slow decline in pulmonary function;

Delay muscle weakness;

- Mendell et al 1989
- Griggs et al 1991
- Angelini et al 1994
- Mesa et al 1991
- Bonifati et al 2000
- Biggar et al 2001

Steroid Treatment and the Development of Scoliosis in Males with Duchenne Muscular Dystrophy

By Benjamin A. Alman, MD, FRCSC, S. Naweed Raza, and W. Douglas Biggar, MD, FRCPC

Investigation performed at the Bloorview MacMillan Children's Centre, The Hospital for Sick Children, and the University of Toronto, Toronto, Ontario, Canada

Steroids in DMD

- 54 boys,
- Age range 7-10 yrs,
- Not randomised,
- Well matched groups,
- 30 received Deflazacort 0.9 mg/kg/day,
- 24 controls

TABLE I Characteristics of the Two Groups at Baseline

	Treatment Group	Control Group
No. of patients	30	24
Pulmonary function* *(% predicted forced vital capacity)*	85 ± 12	85 ± 15
Weight* *(kg)*	21 ± 6	21 ± 3
Age**(yr)*	8.5 ± 3.8	8.9 ± 3.9

*The values are given as the mean and standard deviation.

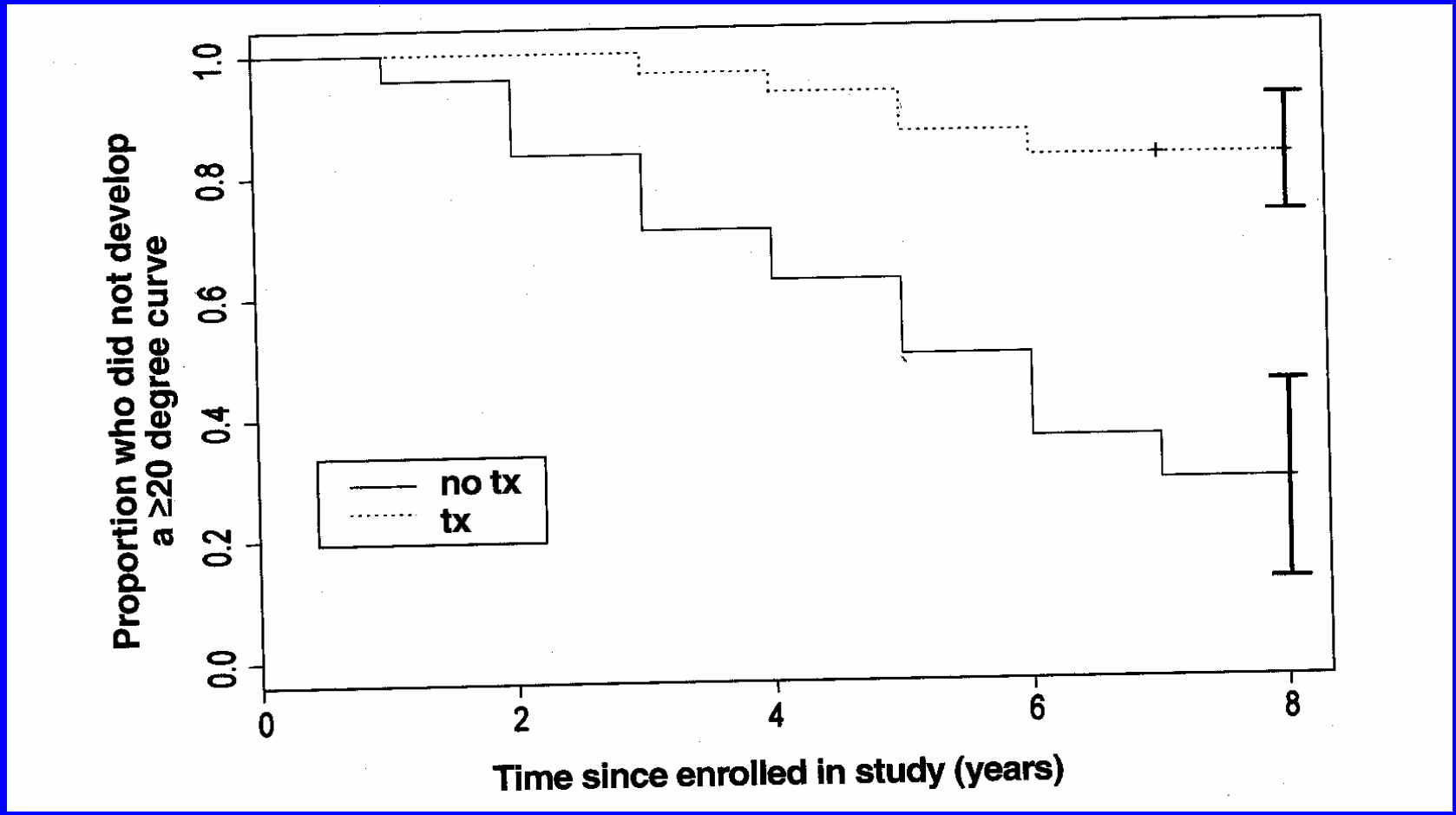
Steroids in DMD- results

TABLE II Comparison of the Two Groups at the Time of Latest Follow-up

	Treatment Group	Control Group
Age stopped walking* *(yr)*	12.3 ± 2.7	9.8 ± 1.8
Age stopped climbing stairs* *(yr)*	10.9 ± 1.9	9.2 ± 1.0
Pulmonary function* *(% predicted forced vital capacity)*	72 ± 11	35 ± 10
Age at op. (for patients who underwent spinal fusion and instrumentation)*	12.9 ± 2.4	15.1 ± 2.0

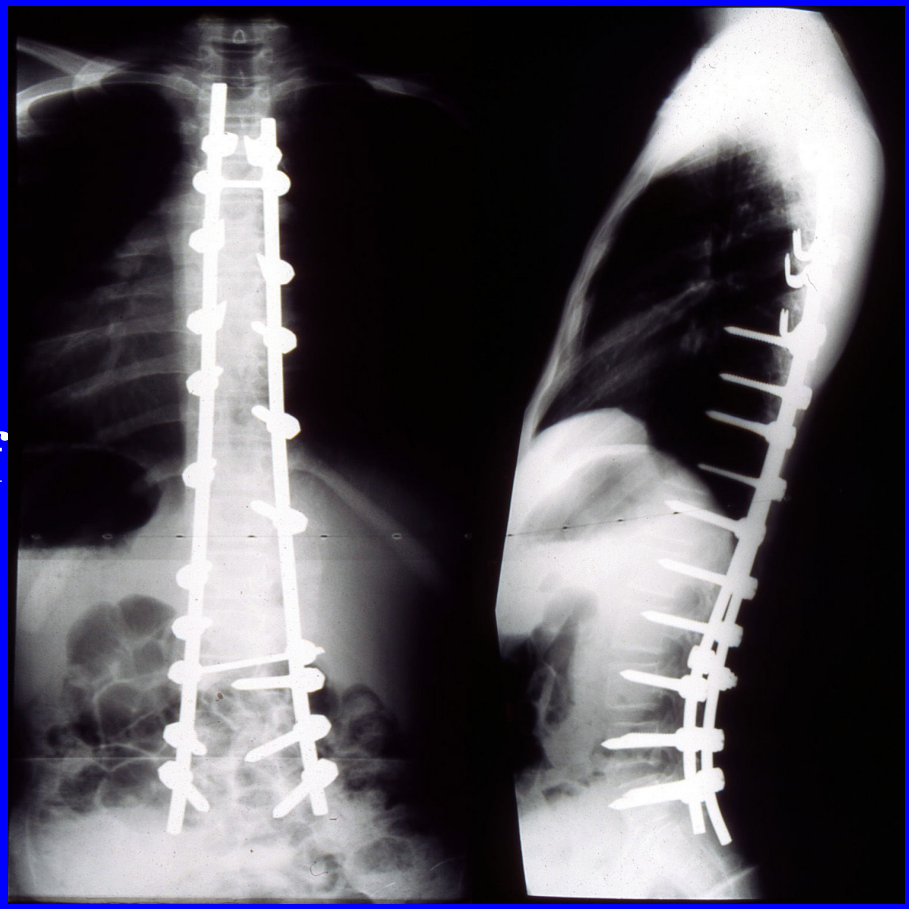
*The values are given as the mean and standard deviation.

Kaplan-Meier Curve for proportion of boys not developing a 20 degree scoliosis/ year



Gene therapy to treat DMD

- **DMD is due to mutation in the dystrophin gene;**
- **Dystrophin maintains integrity of myocytes**



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Correction of Neuromuscular Scoliosis in Patients With Preexisting Respiratory Failure

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Jwalant S. Mehta, MS(Orth), MCh(Orth), FRCS(Orth),* Michael J. Gibson, FRCS,*
K. Bushby, MD, FRCP,† and R. Bullock, MA, FRCP, FRCA‡

Scoliosis Surgery in Myopathic Patients With Pre-op Respiratory Failure

- 8 patients- 6 boys & 2 girls
- Mean age12 yrs (8-15)
- Mean follow up 48 mths (12-80)
- 4 Multicore myopathy, 3 congenital muscular dystrophy, 1 acyl-COA dehydrogenase deficiency

NOCTURNAL VENTILATION

INDICATIONS FOR VENTILATION:

- 7 patients, early symptoms of respiratory failure:
  repeated waking at night,
  daytime somnolence,
  general malaise,
  repeated chest infections

Diagnosis confirmed by overnight pulse oxymetry.

- 1 patient admitted in severe respiratory failure.


BiPAP S/T 30

NOCTURNAL VENTILATION

- TECHNIQUE:

BIPAP (Biphasic positive airway pressure ventilation) overnight whilst the patient is asleep via either a mask or nasal pillows.

- FOLLOW UP:

3-6 monthly at the muscle clinic with PFTs.

Yearly overnight oxymetry.

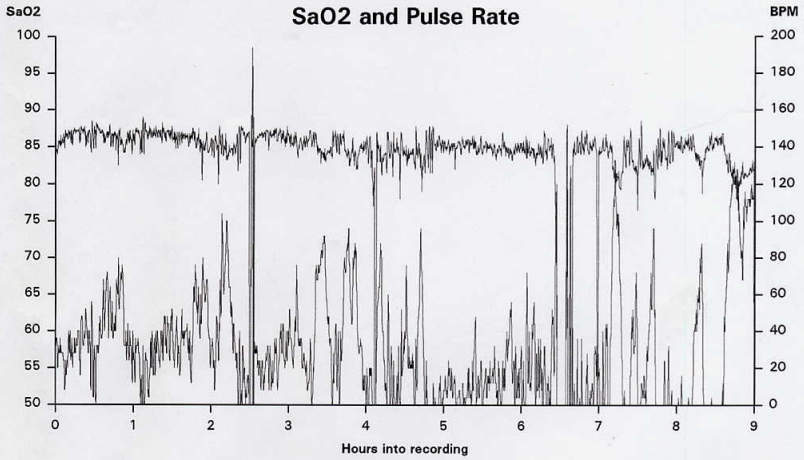
- RESULTS:

BIPAP is effective in reversing episodes of desaturation during sleep at night.

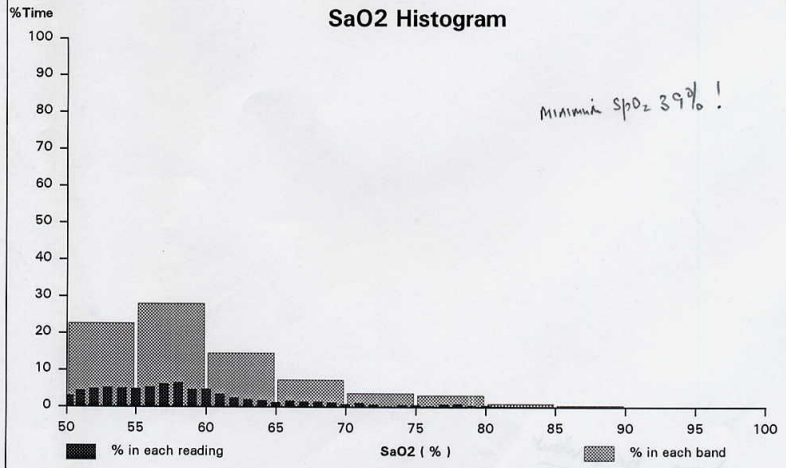
PULSE OXIMETER RECORDING byox0002.dat

Patient Name	**John Padley**
Recording Started	**30/08/1996 00:00**
Recording Length	**08:00**
Time at less than 90% SaO2	**07:47:24 (97.3%)**

SaO2 and Pulse Rate



SaO2 Histogram



Minimum SpO2 39% !

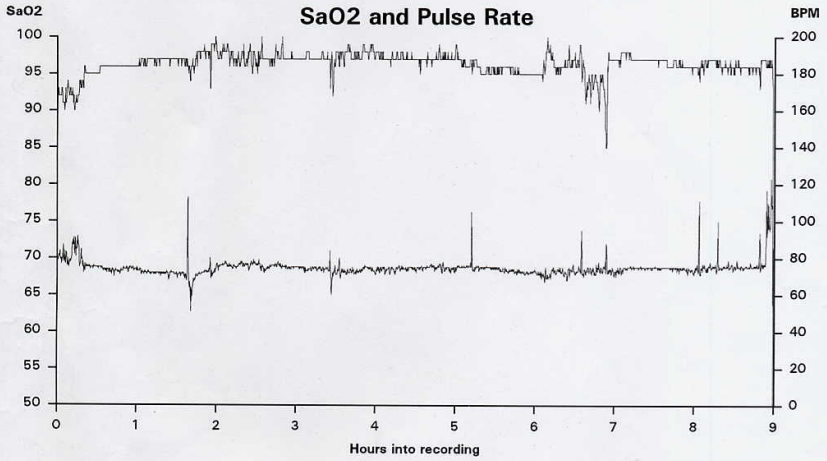
Record prepared on : 30/08/96

andyc@rmpd-ngh.demon.co.uk

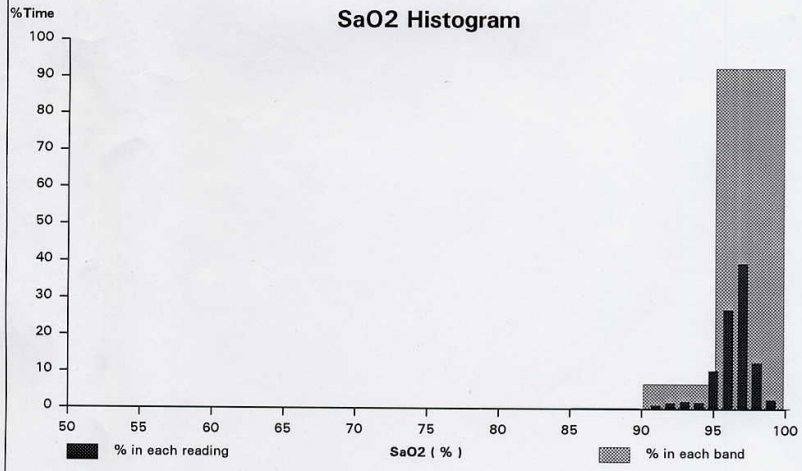
PULSE OXIMETER RECORDING byox0003.dat

Patient Name	**John Padley**
Recording Started	**02/09/1996 00:00**
Recording Length	**08:00**
Time at less than 90% SaO2	**00:01:12 (0.2%)**

SaO2 and Pulse Rate



SaO2 Histogram

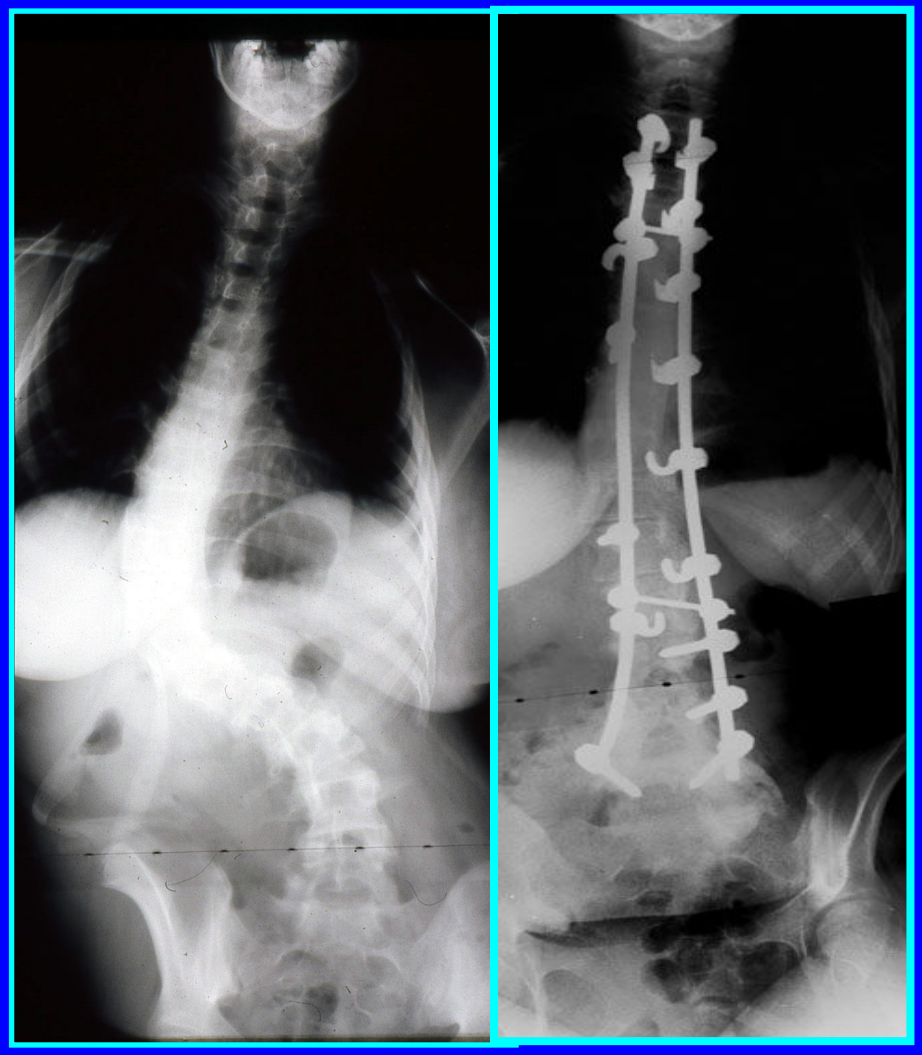


Record prepared on : 02/09/96

andyc@rmpd-ngh.demon.co.uk

Surgery in Myopathic Patients with Pre-op Respiratory Failure

- ITU stay 2-5 days (2.7)
- Hospital stay 10-21 days (12.2)
- VC pre 20% post 18%
- Blood loss 1450 ml (500-2600 ml)
- Cobb angle pre-op 70.2 post-op 32



FUNCTIONAL STATUS

Pre- op:

4 Patients were ambulant

5 patients were independent sitters

Post-op:

All patients maintained their pre-op level of function.

Treatment of Scoliosis in Ambulant Myopathic Patient

- Bracing Does not Work
- Surgery to maintain spinal balance


AP ERECT
L


LT
SITTING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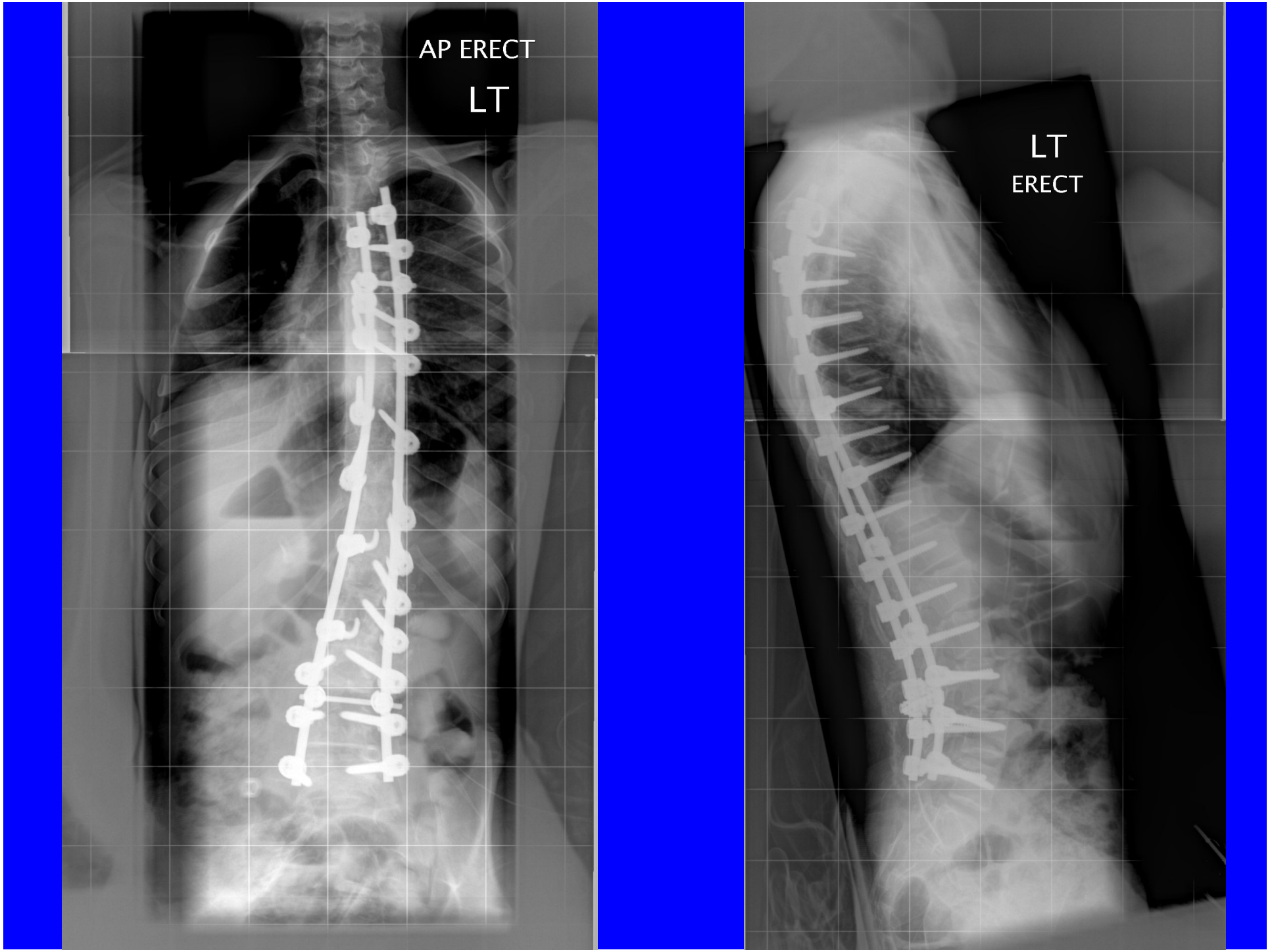
SUPINE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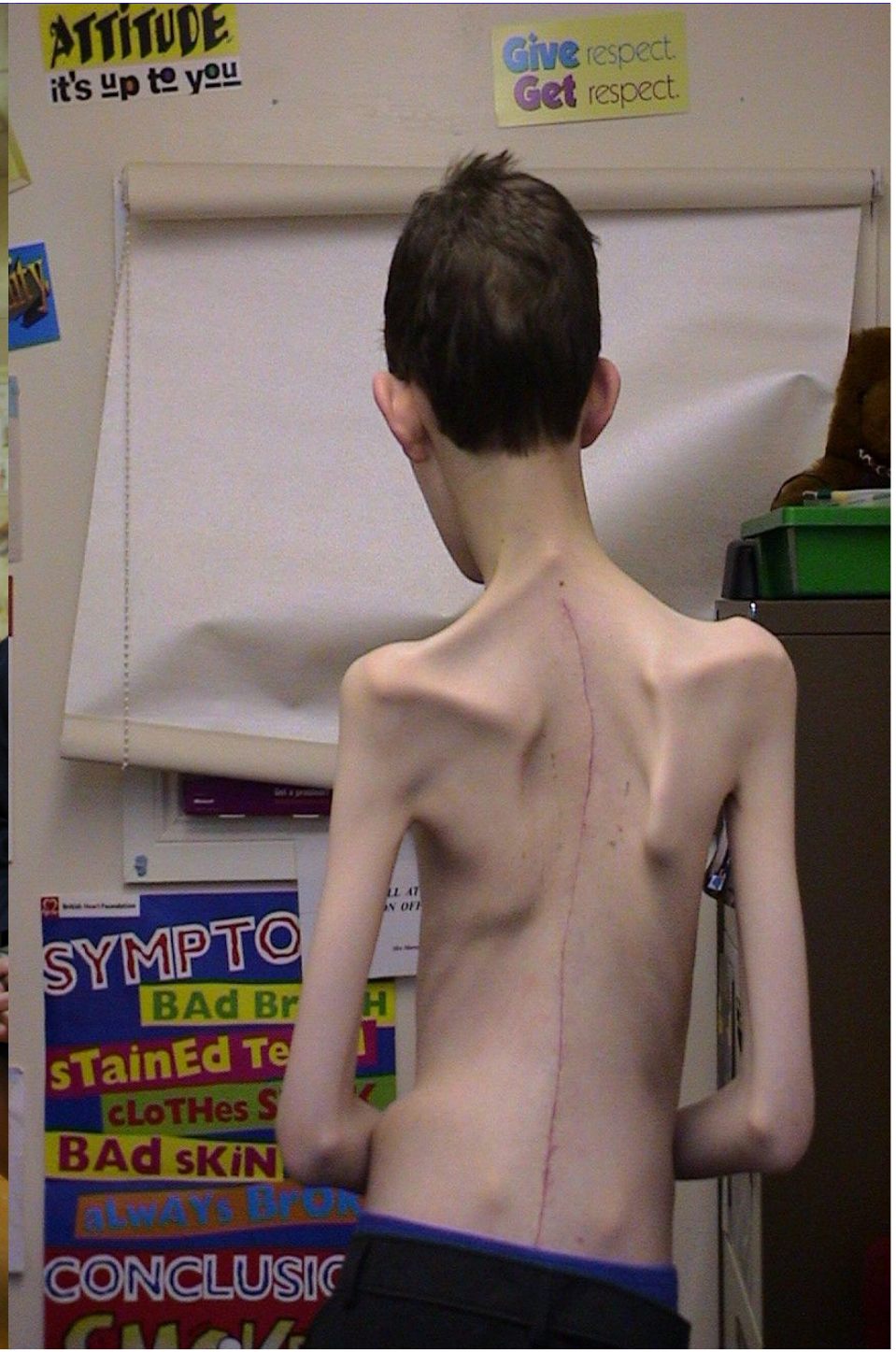

ATTITUDE
it's up to you
Give respect.
Get respect.
SYMPTO
BAd Br H
sTainEd Te
cLoTHes S
BAd sKiN
aLWAYs Bro
CONCLUSIO

Scoliosis in Cerebral Palsy

Incidence of scoliosis in CP 25.6% (Samilson & Bechard)

- 232 had scoliosis out of 906 patients;

  22 ambulant.

  41 independent sitters.

  169 bed care .

Incidence of Scoliosis in CP

- 75% of severely affected quadriplegic patients have a scoliosis.
- 6-10 % of ambulatory diplegics & hemiplegics have a scoliosis.

Madigan and Wallace (1981)

Classification

- Classified by Neurology
  - Spastic
  - Athetoid
  - Ataxic
- Classified by Involvement
  - Hemi-plegia
  - Diplegia
  - Total Body Involvement.

Natural History of Scoliosis in CP

- Madigan et al - Scoliosis in the Institutionalised CP population. **Spine 1981**
- Thometz et al – Progression of scoliosis after skeletal maturity in Institutionalised adults who have CP. **JBJS 1988**
- Majd E et al – Natural History of scoliosis in the institutionalised adult CP population. **Spine 1997**
- Saito et al – Natural History of scoliosis in spastic CP. **Lancet 1998**

Natural History of Scoliosis in CP

- Mean age of onset 10 yrs
- Curves of > 40 under 15 yrs will progress
- Quad, Thoracolumbar curves and Severe neurology highest rates of progression
- Progress after maturity- 25% stop at 22 yrs
- Average age of Risser 5 was 23 years
- 30 yrs survival rate 87%.

  95% in hemi or mono

  83% in quads

1992

Cobb = 90

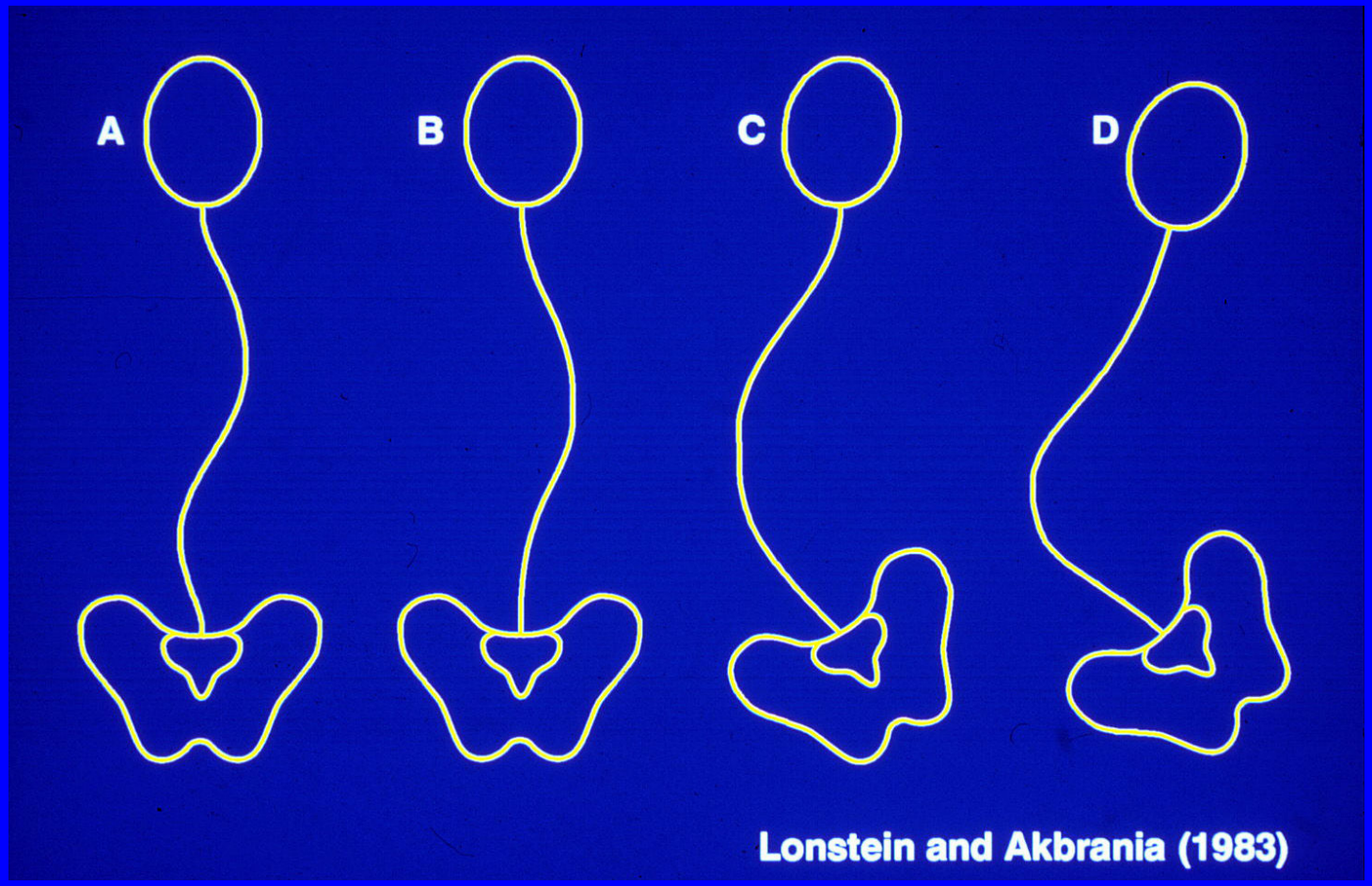
1993

Cobb = 110

Natural History of Scoliosis in CP

- Progression of curve
  - Poor sitting balance
  - Poor upper limb function (Boachie-Adjeli JBJS 1989)
  - Pressure sores and pain
  - Respiratory compromise
  - Bowel dysfunction
  - Discomfort
- Caregiver input
  - Longer time
  - More intense effort

Curve Patterns Patients with Scoliosis secondary to C.P.

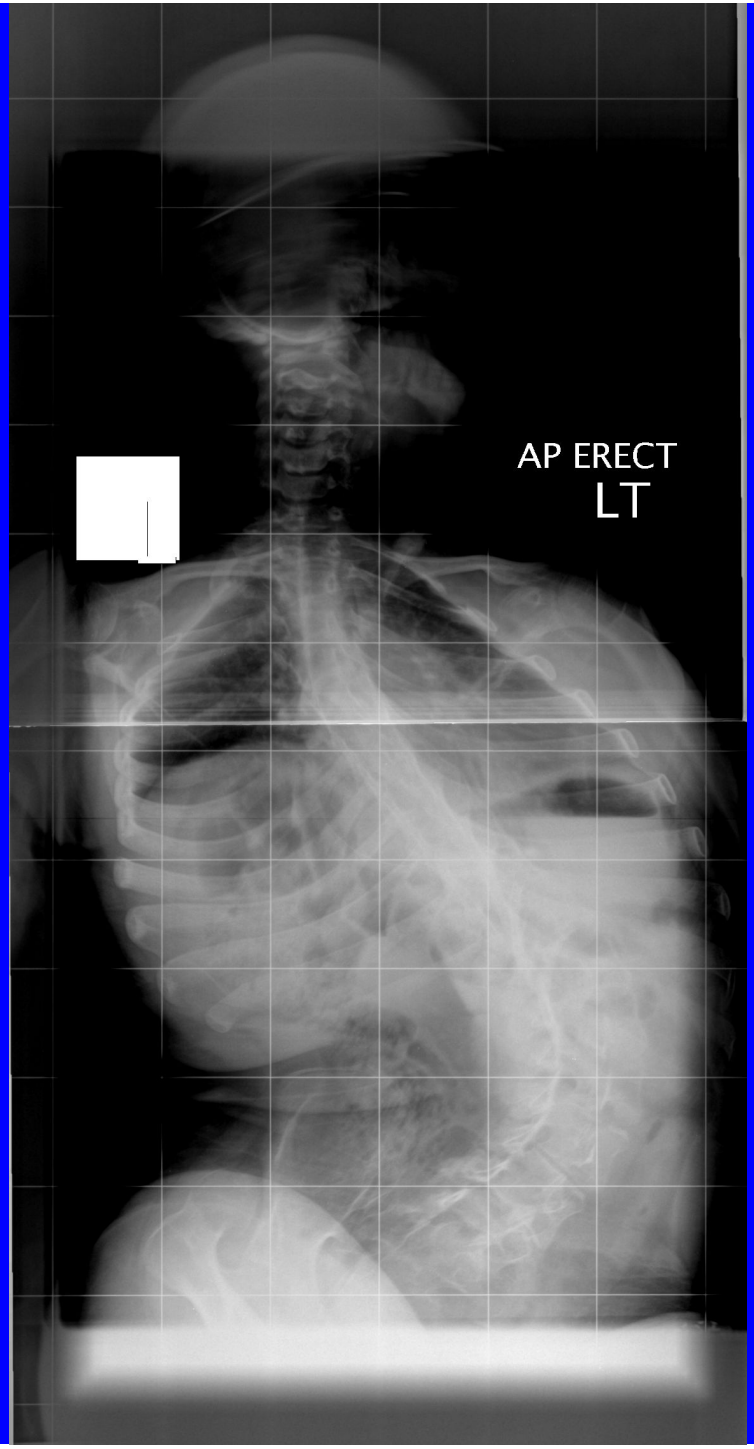


Treatment of scoliosis in CP

- Lonstein type A&B- Level pelvis; good function ambulant or independent sitters; Instrument and fuse to L5.
- Lonstein type C&D-Largest group; oblique pelvis; severely disabled little independent function; ? Candidates for surgery; Posterior instrumented fusion to pelvis with anterior release for stiff curves.

Scoliosis in Severe Spastic Quad

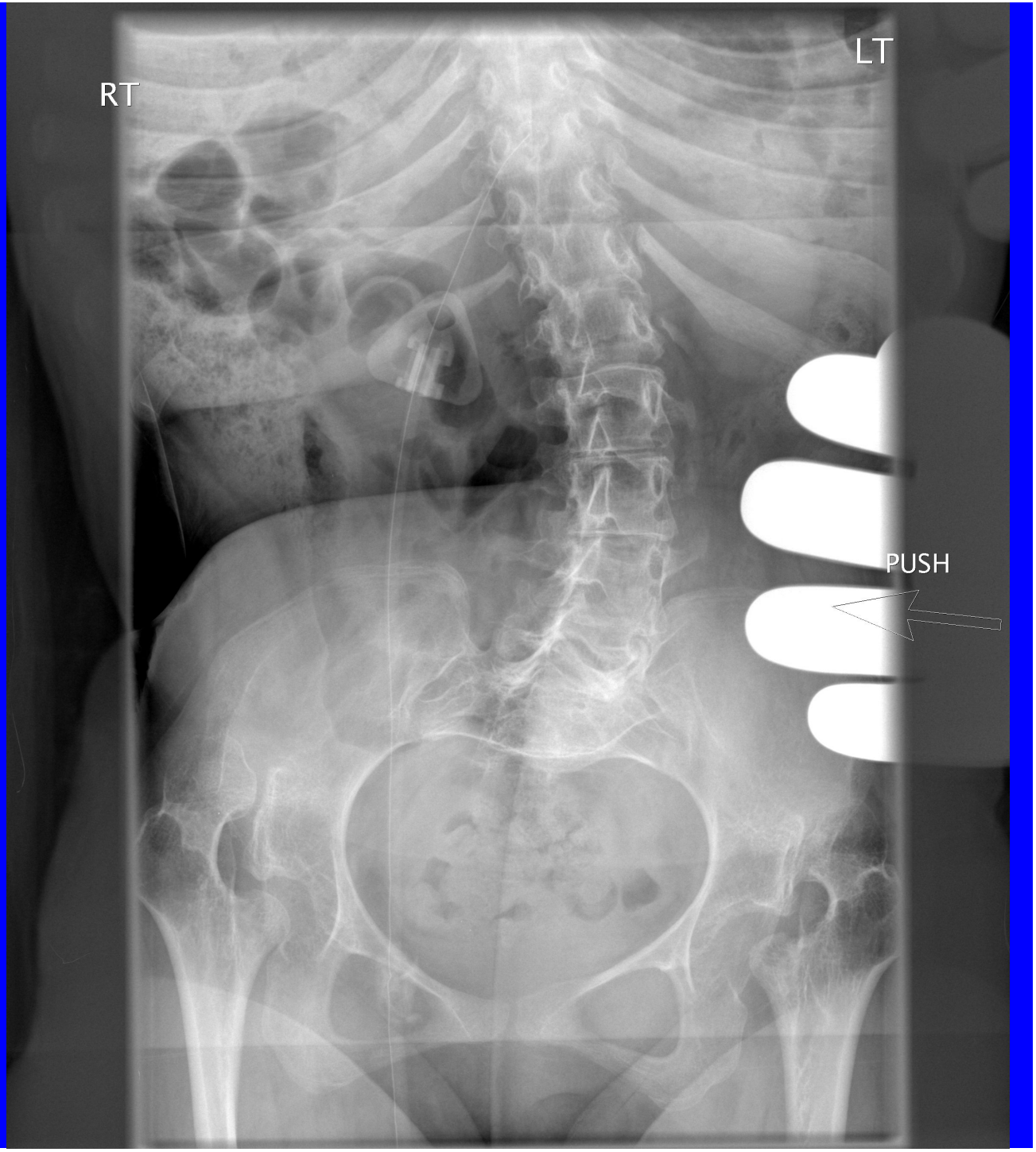
- ? Treatment



Spastic Quad

Surgery

- Post alone?
- Ant + Post?
- 1 or 2 stage?

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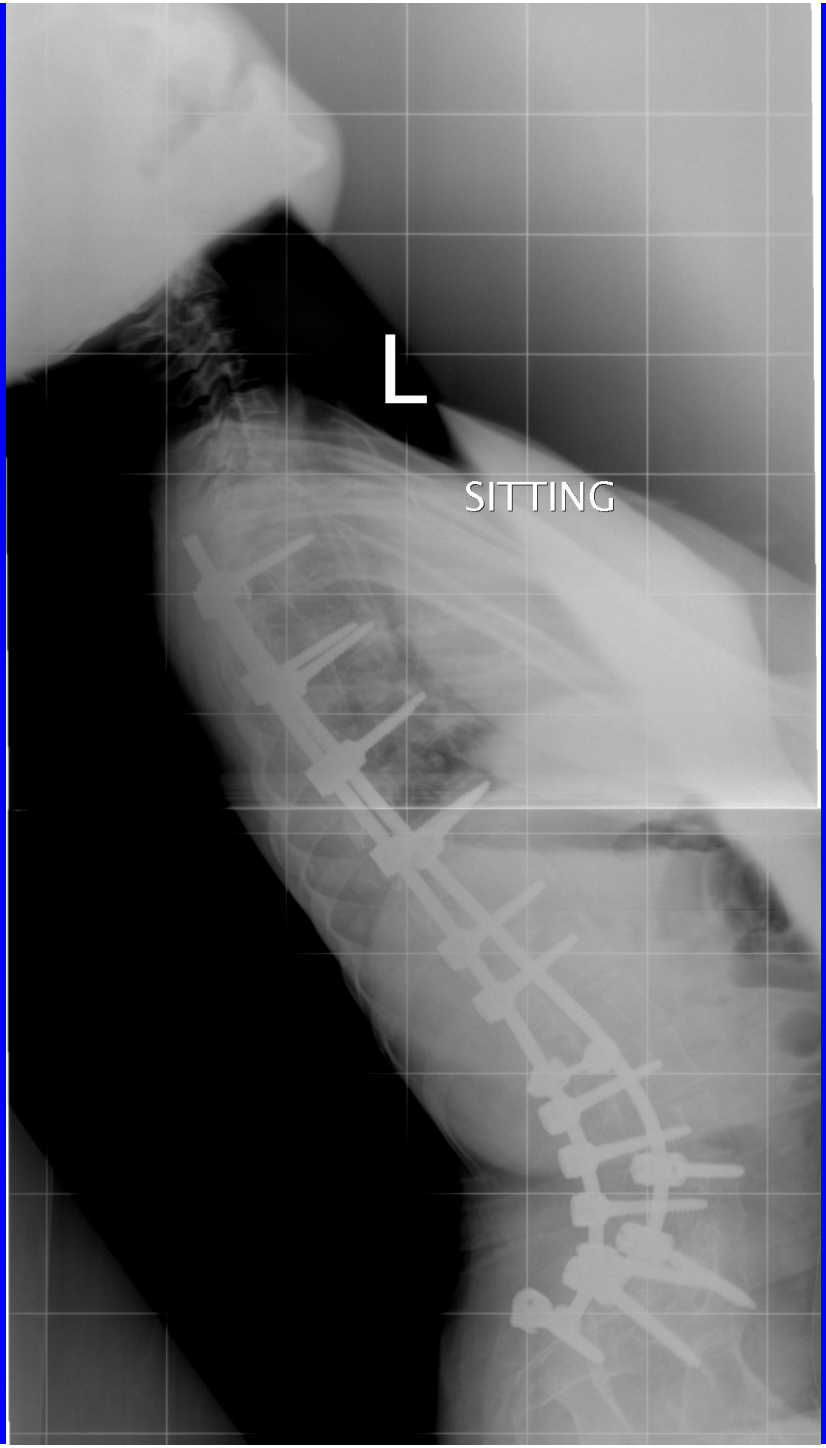
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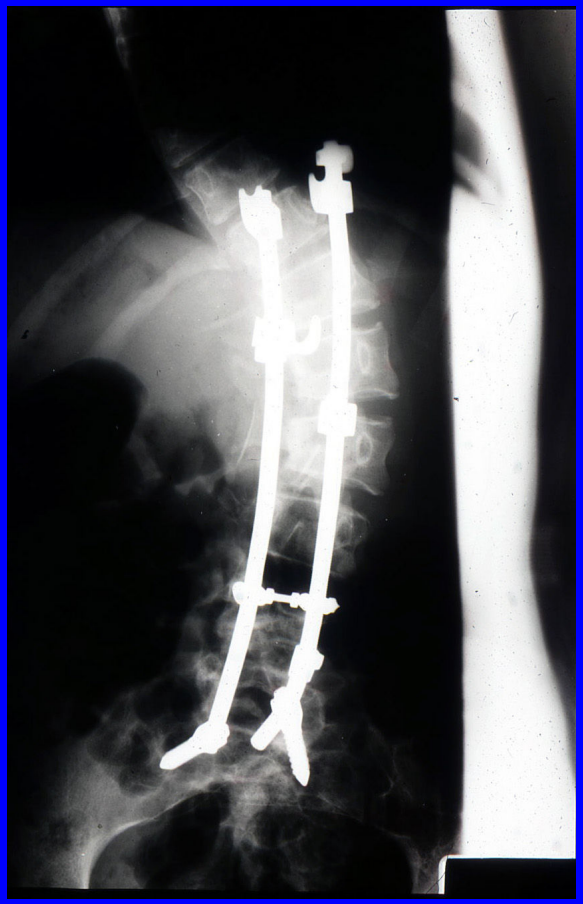
Complications of surgery in neuromuscular scoliosis

High complication rate;

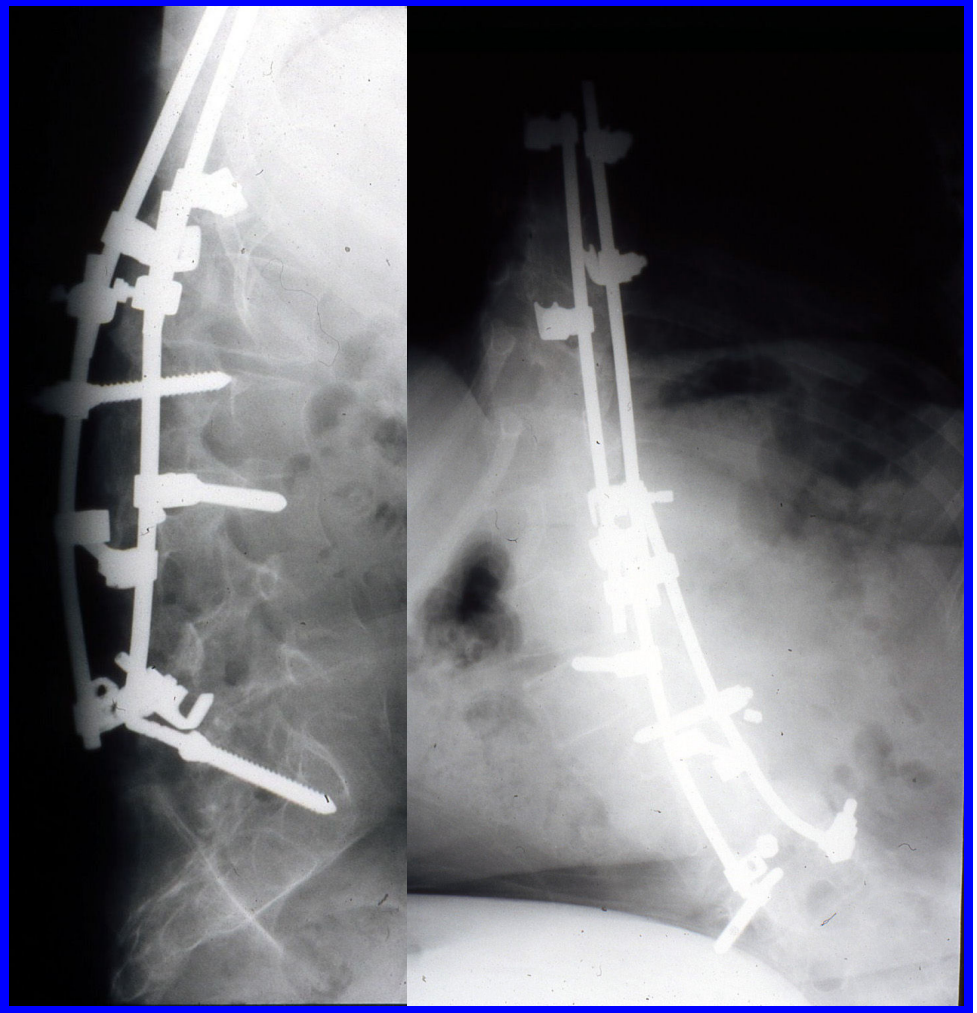
- 70% in early series
- Intra operative bleeding!
- Failure of fixation
- 10% late infection



Complications- Fixation Failure

Neuromuscular Scoliosis

- Careful planning
- Rigid fixation
- Immediate mobilisation
- “One chance”



Neuromuscular Scoliosis

- Identify Candidates for surgery Early
- Operate Before Curve is Stiff
- Long Posterior Instrumentation +/- Pelvis
- Surgery improves Quality of Life & Longevity