

# SCOLIOSIS IN RETT SYNDROME

# INCIDENCE

- Overall incidence-approx 1 in 10,000 to 15,000 live female births
- Reported incidence of scoliosis varies between 30-100%.
- Variation probably related to diagnostic accuracy, patient age, and condition severity in various series.
- Incidence of scoliosis probably about 60%

# AGE OF ONSET

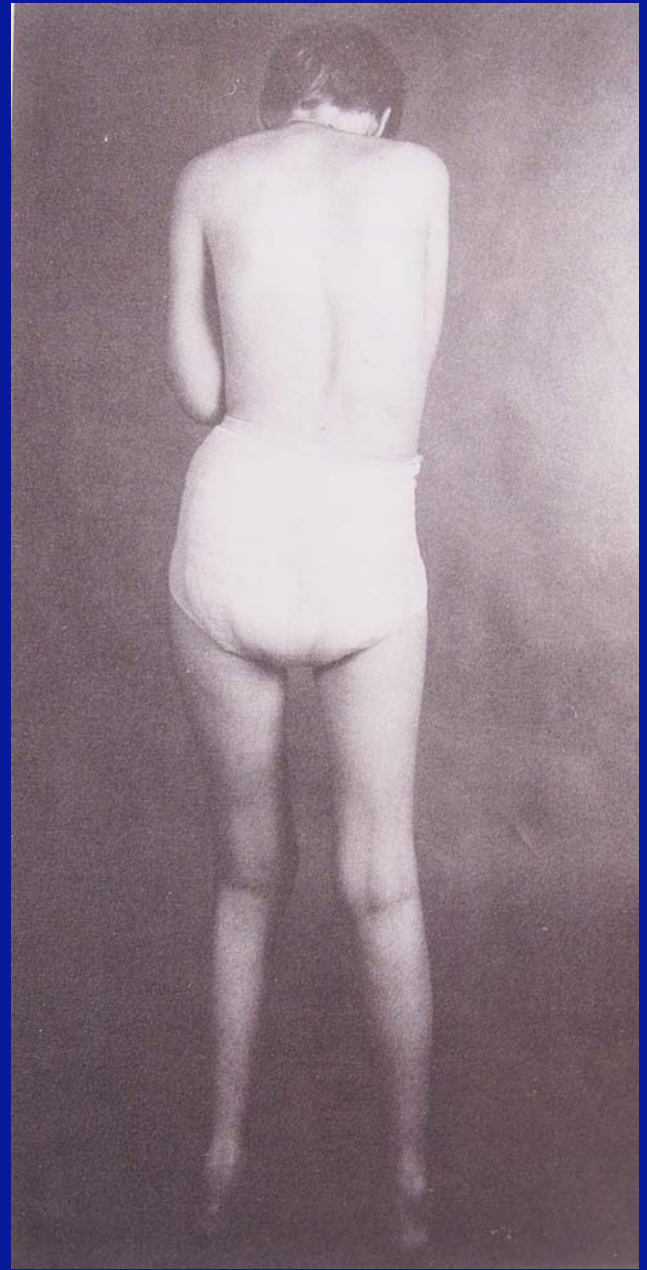
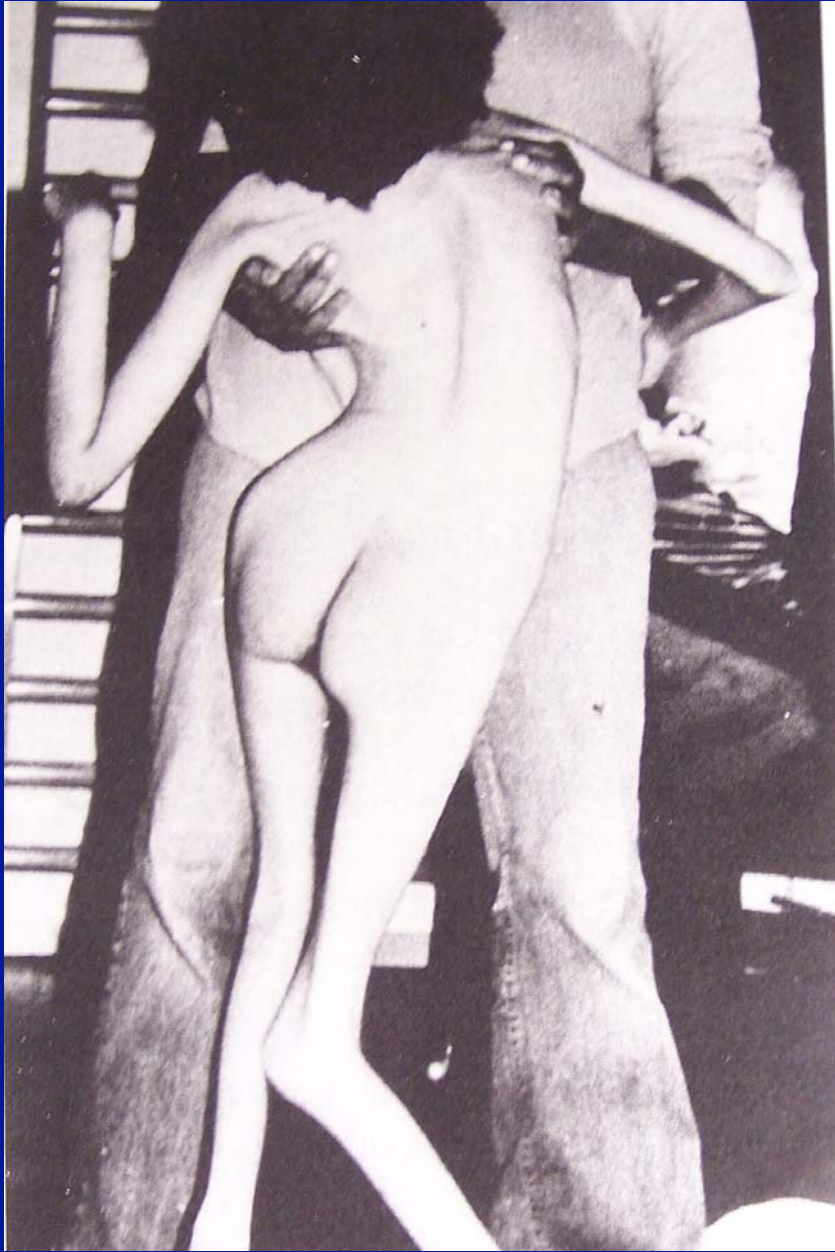
- Onset earlier than most scoliotic conditions.
- 3% before clear diagnosis.
- Often by age 4.
- May begin while still ambulant.
- Depends on stage and severity of condition.

# CURVE PROGRESSION

- May be rapidly progressive.

# CURVE TYPES

- Majority probably have classical long C shape neuromuscular curve.
- Some patients have more idiopathic type double or thoracic curves.



# PROBLEMS OF SCOLIOSIS

- Loss of trunk balance causing difficulties walking or more commonly sitting.
- If curve is severe ( $>70^\circ$ ), may cause respiratory or feeding problems.
- Pelvic obliquity.





# CONSERVATIVE TREATMENT

- BRACING - may slow curve progression.
  - best for curves  $< 30^\circ$  □
  - results variable.
- . WHEELCHAIR MODIFICATION.- to assist with seating balance
- . MONITORING - every 6 months.
  - more frequently in early advanced neurological impairment



# INDICATIONS FOR SURGERY

- Curve progression.
- Loss of trunk balance.
- Surgery is better undertaken before the curve is severe ( $<60^\circ$ ) and pelvic obliquity occurs.



# AIMS OF SURGERY

- Control curve progression.
- Restore trunk and sitting balance.

# SURGERY

- Need to be performed in major centre.
- Need intensive postop medical supervision and access to high dependancy or intensive care.

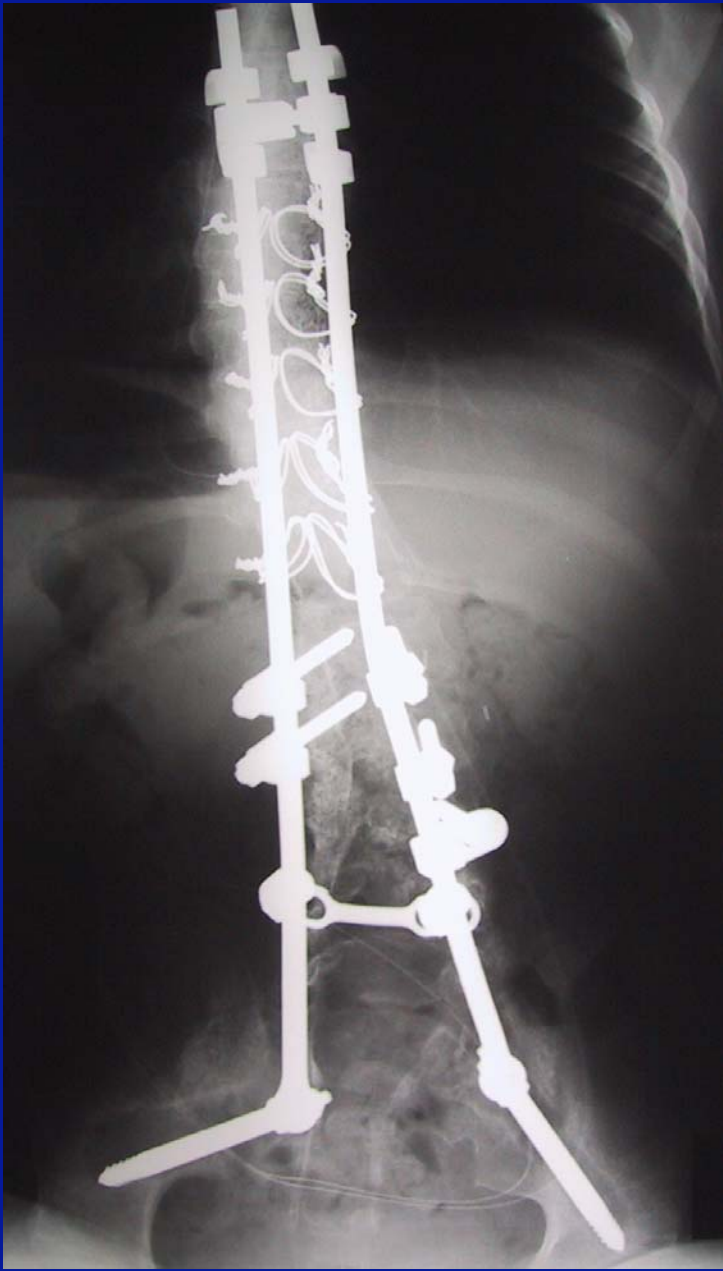
# SURGERY

- Posterior or anterior/ posterior.
- Anterior required for
  - young child.
  - severe deformity.
  - stiff deformity.
  - pelvic obliquity.

# LENGTH OF FUSION

- Entire curve needs to be fused. Longer rather shorter as condition progressive.
- In milder more flexible curves can stop fusion at L4 or L5.
- If severe stiff pelvic obliquity, fixation to pelvis may be required. This is a much bigger procedure with risk of complications.





# SURGICAL DIFFICULTIES

- Patients nutritional status.- ? Need for preop augmented feeding.
- Low body weight - low blood volume
- Wound healing problems- infection or breakdown - plastic surgeon
- Osteoporosis.\_ fixation problems
- Respiratory problems.Unable to cough or deep breath at request.
- Incontinence
- Epilepsy
- Familial distress.
- Patient distress.

# SURGICAL BENEFITS

- Adequate sitting balance
  - patient much happier
  - frees up carer
- Improved nutrition - less GIT reflux.
- Improved respiratory function - decreased chest infections.
- Effect on walking variable
  - long recovery time
  - stiff spine

# SUMMARY

- Scoliosis common and a major cause of difficulties.
- If scoliosis progressive , surgery better considered earlier rather later.
- Surgery is a major undertaking.
- Results frequently very beneficial to patient and carer.