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 (>70º), may cause respiratory or feeding problems.
• Pelvic obliquity.
CONSERVATIVE TREATMENT

• BRACING - may slow curve progression.
  - best for curves < 30°
  - 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°) 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 than 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.