

Dynamic Thoracoplasty Juene Syndrome

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Juene Syndrome

- Familial Asphyxiating Thoracic Dystrophy
- Juene 1955
- Rare $1 / 10^5$ live births
- Autosomal recessive acromelic dwarfism
- Variable severity reflects genetic heterogeneity
- Restrictive lung disease
- Renal / pancreatic disease
- Neonatal Juene Syndrome is fatal in 70 %

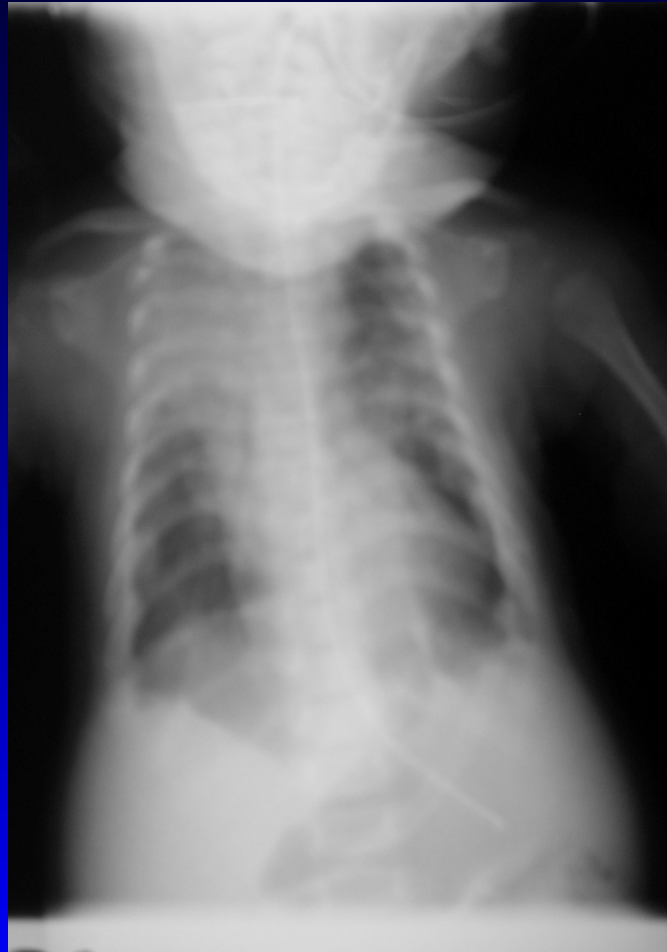
Juene syndrome

Locus at chromosome
15q13

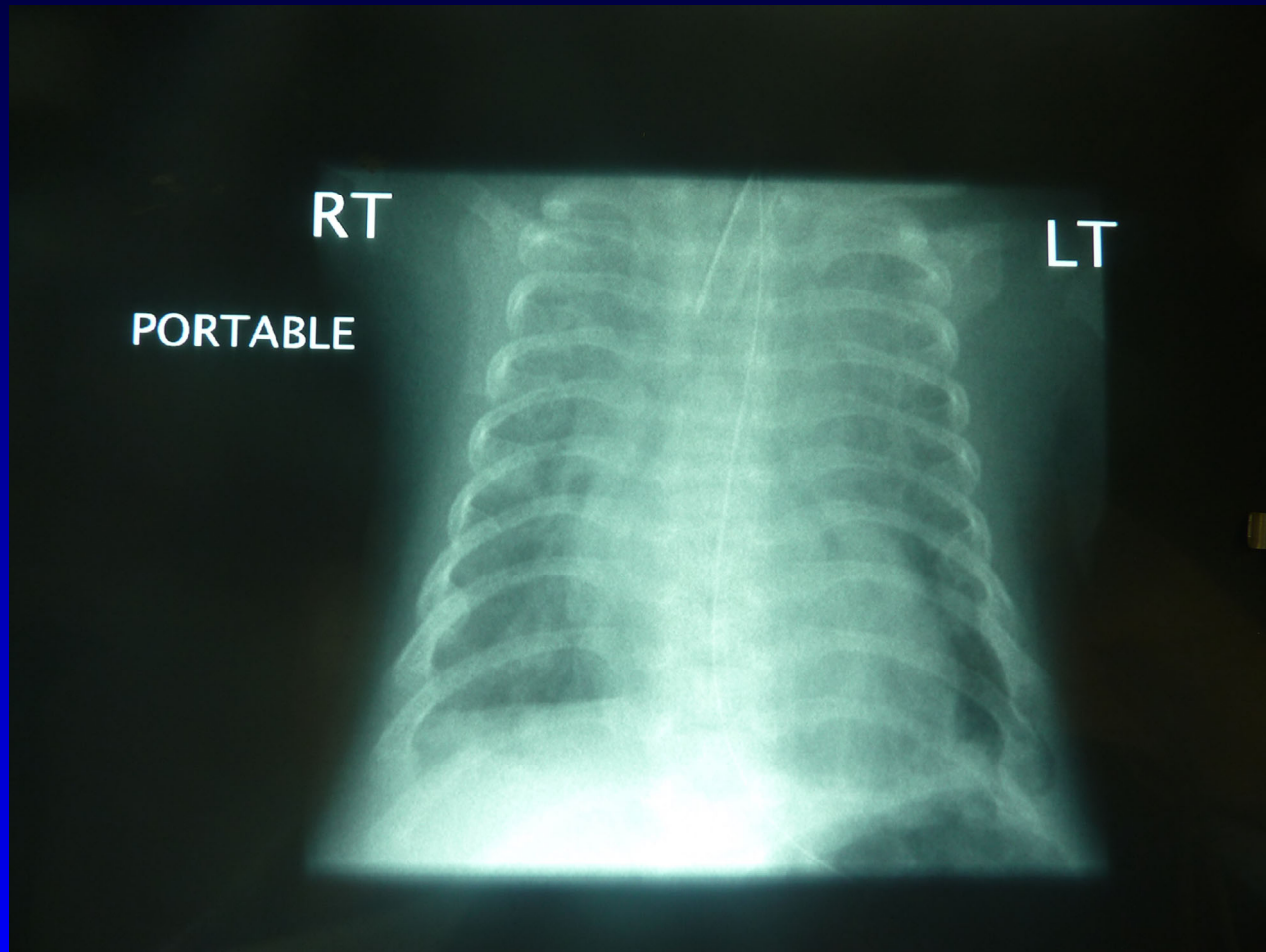
Locus at 3q24-3q26

A chondrodysplasia
linked to a defect in
primary cilia function
or intraflagellar
transport

Nat Genet Jun 2007

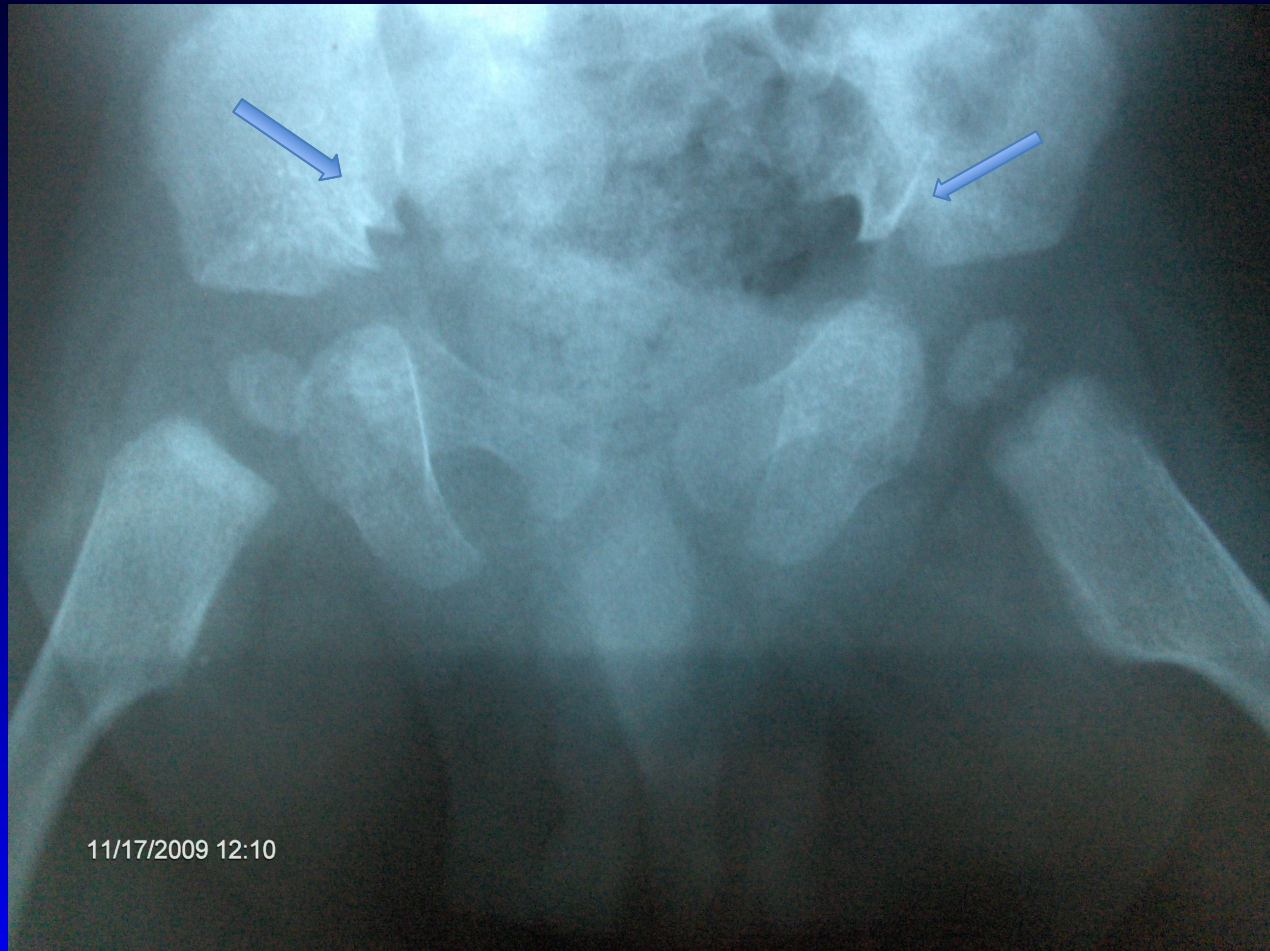


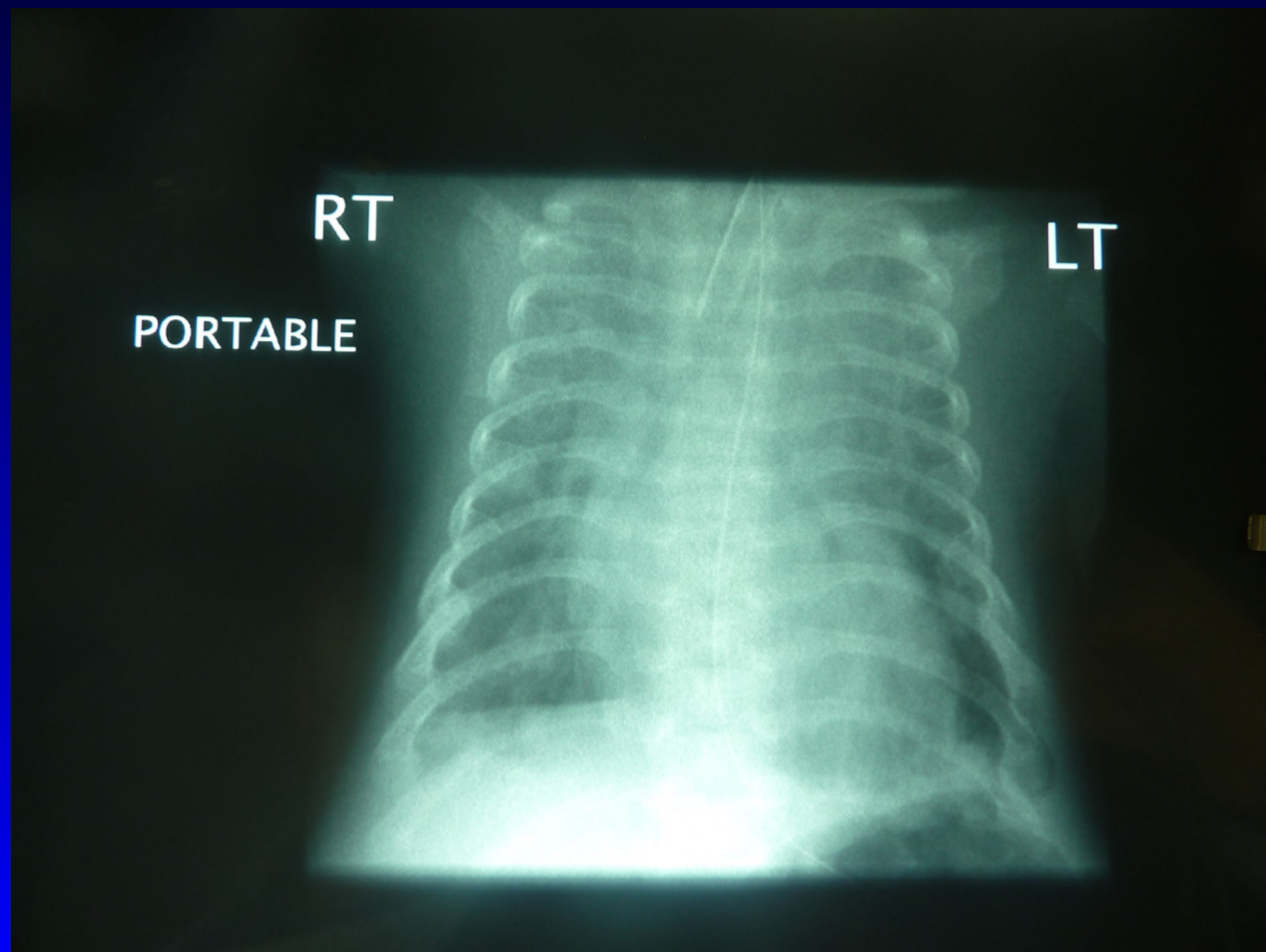
Juene syndrome



Juene syndrome

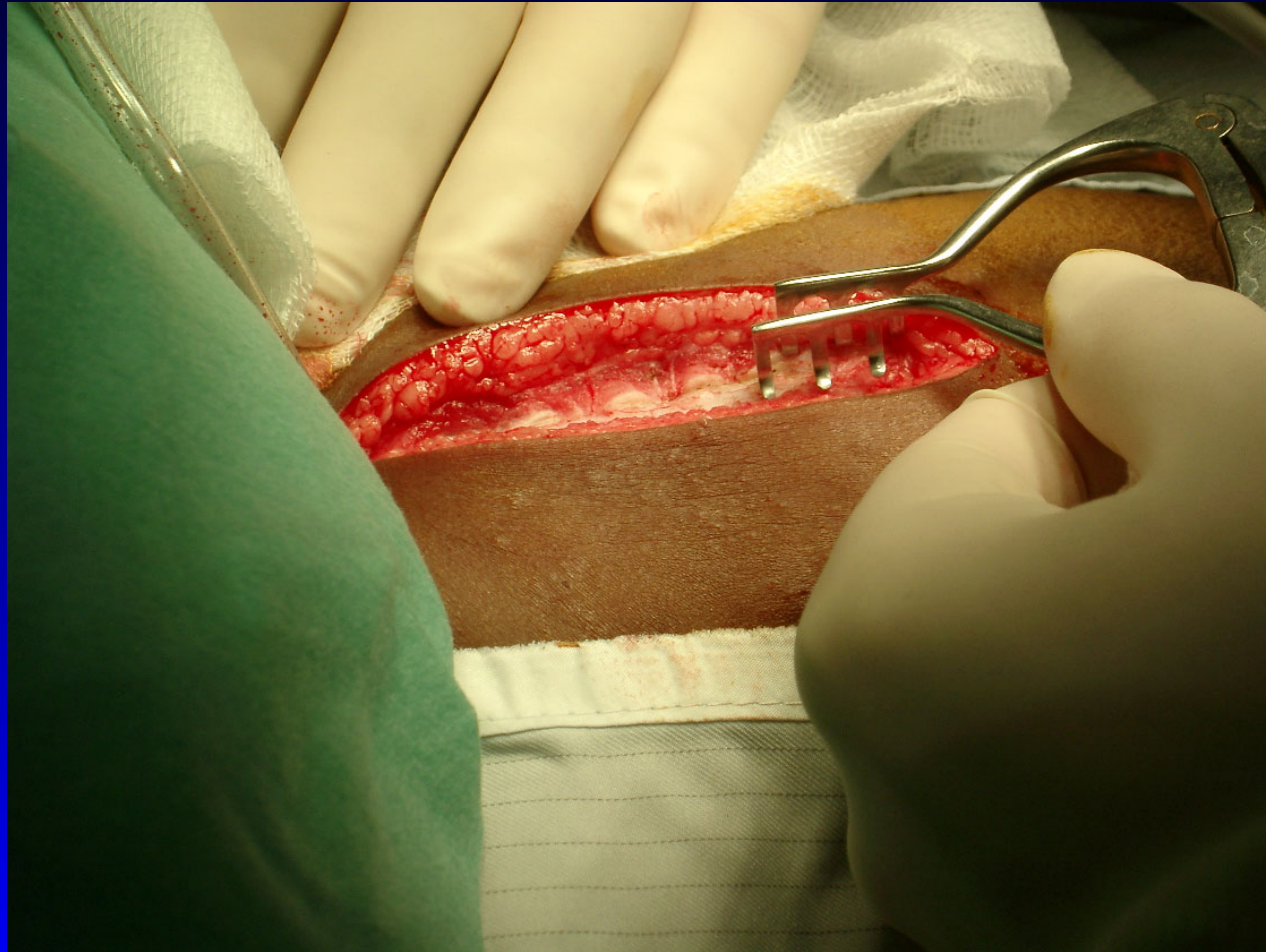
sciatic notch bone spikes



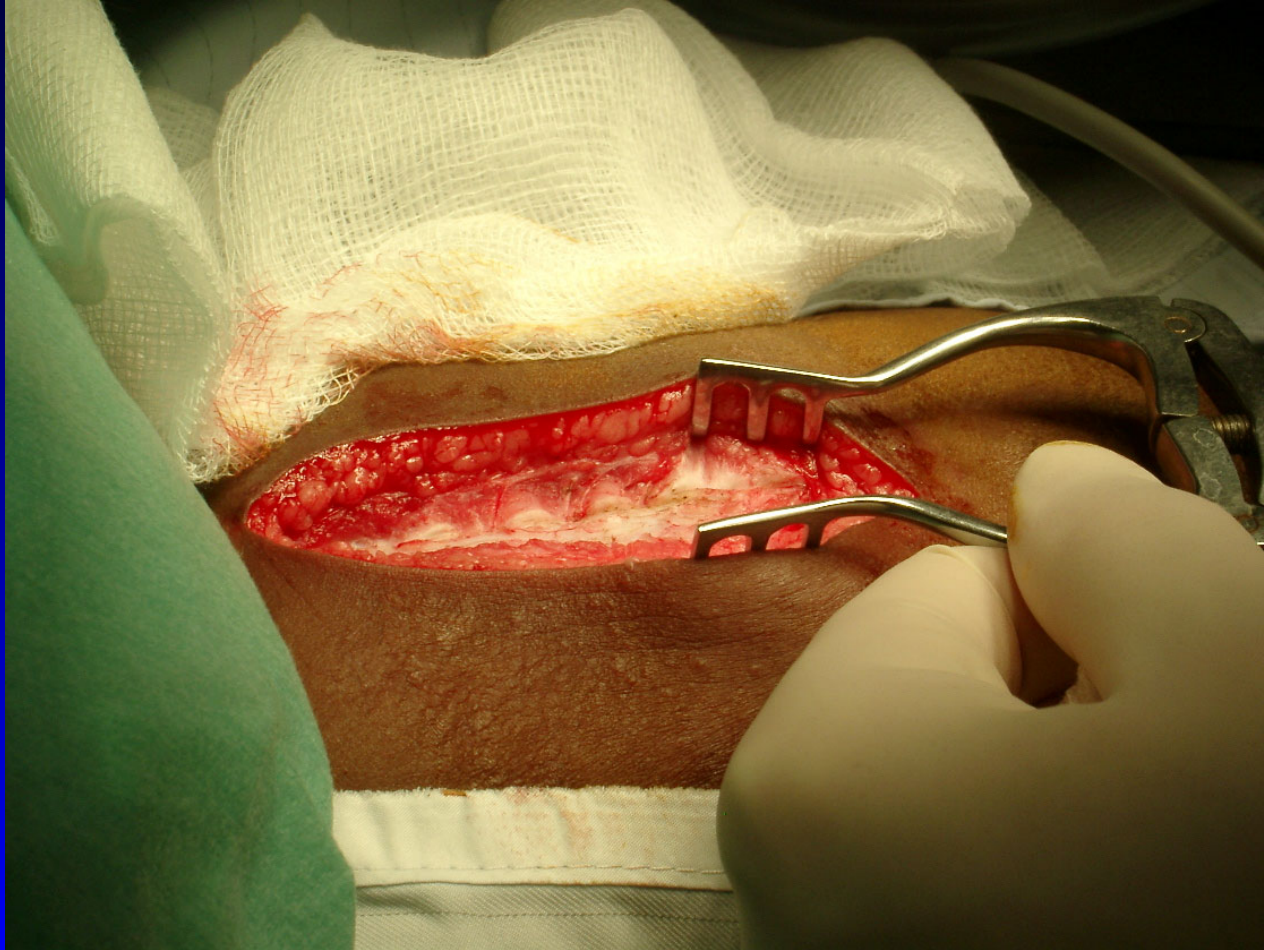




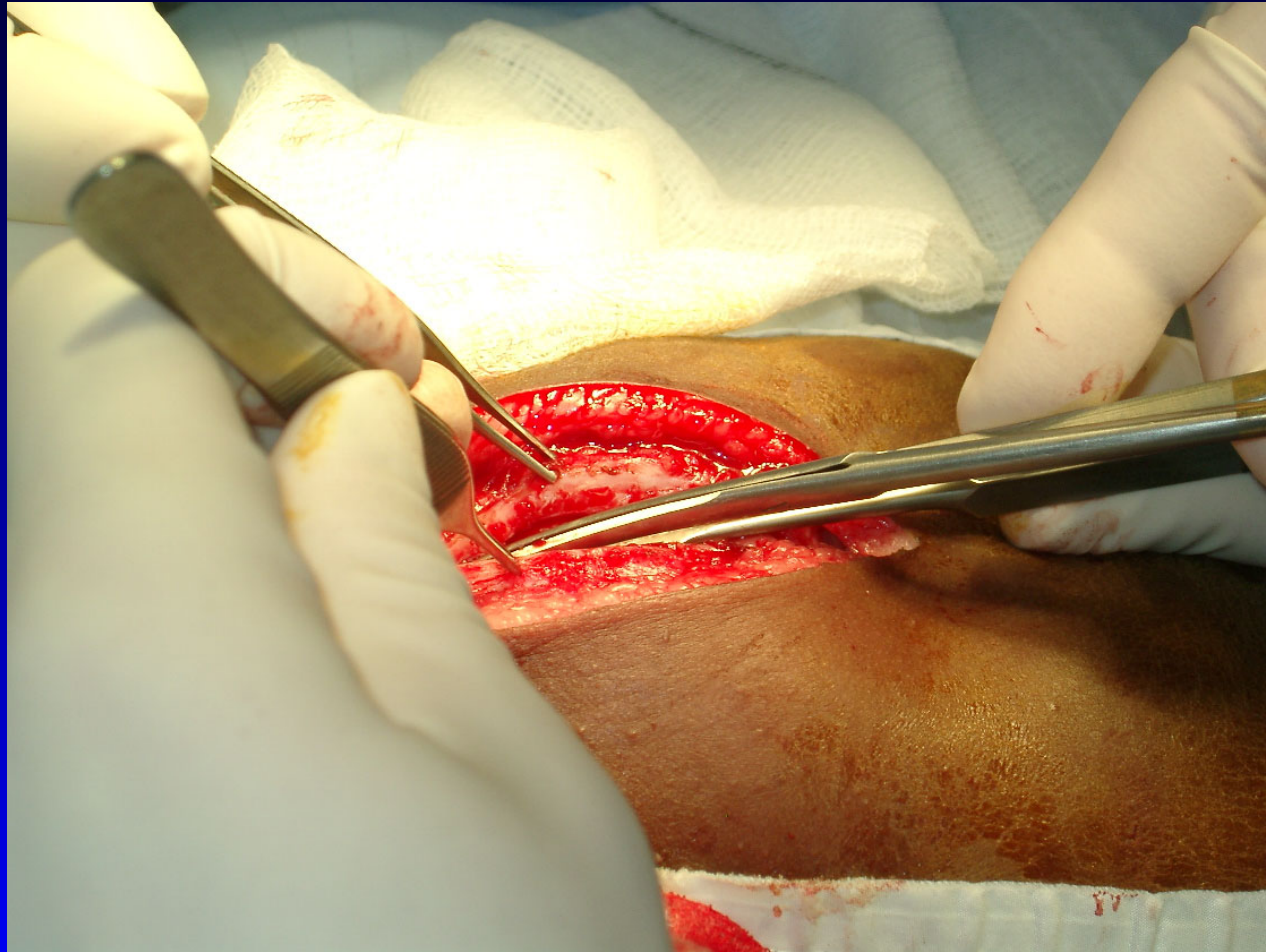
sternotomy

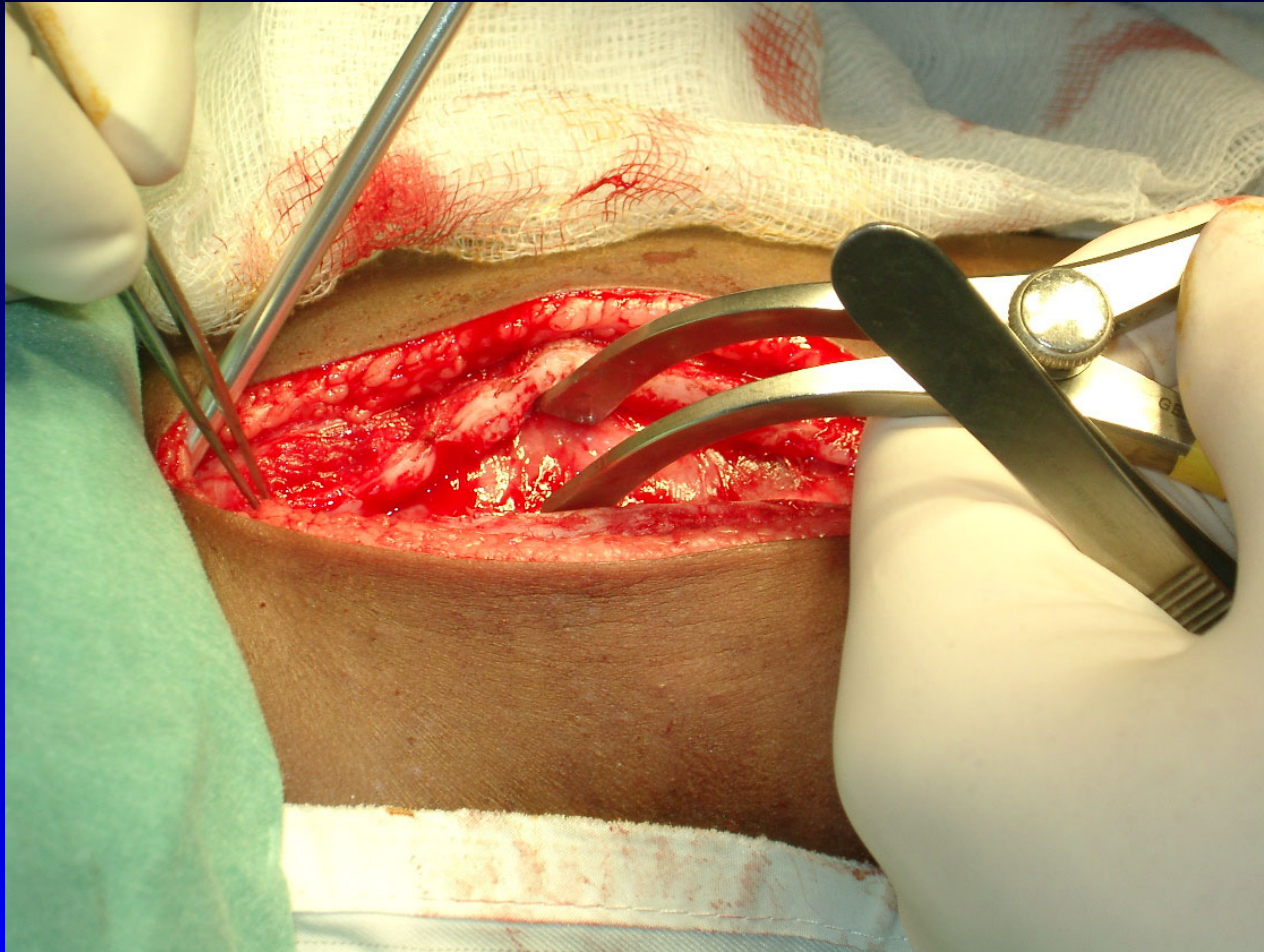


sternotomy

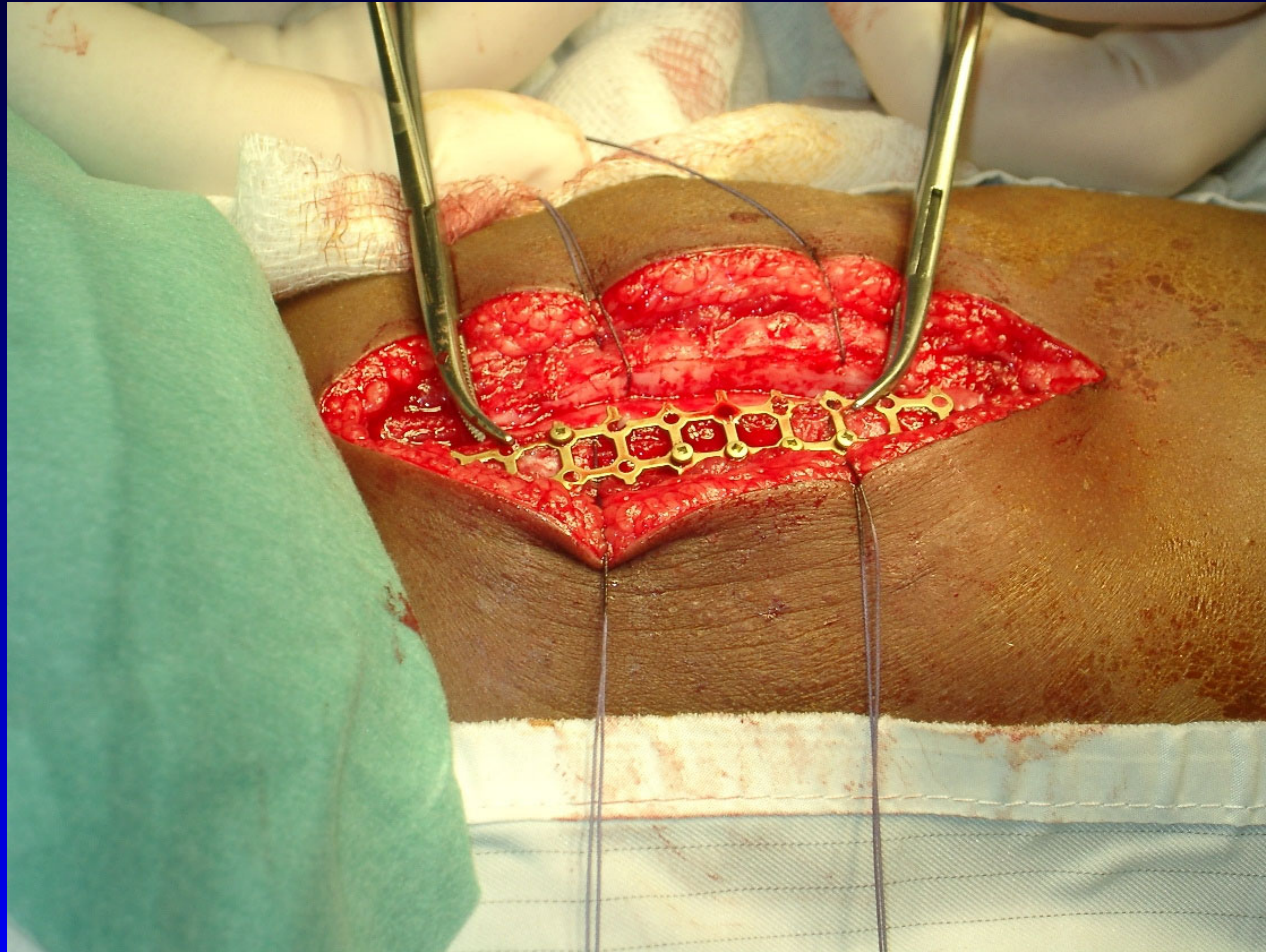


sternotomy

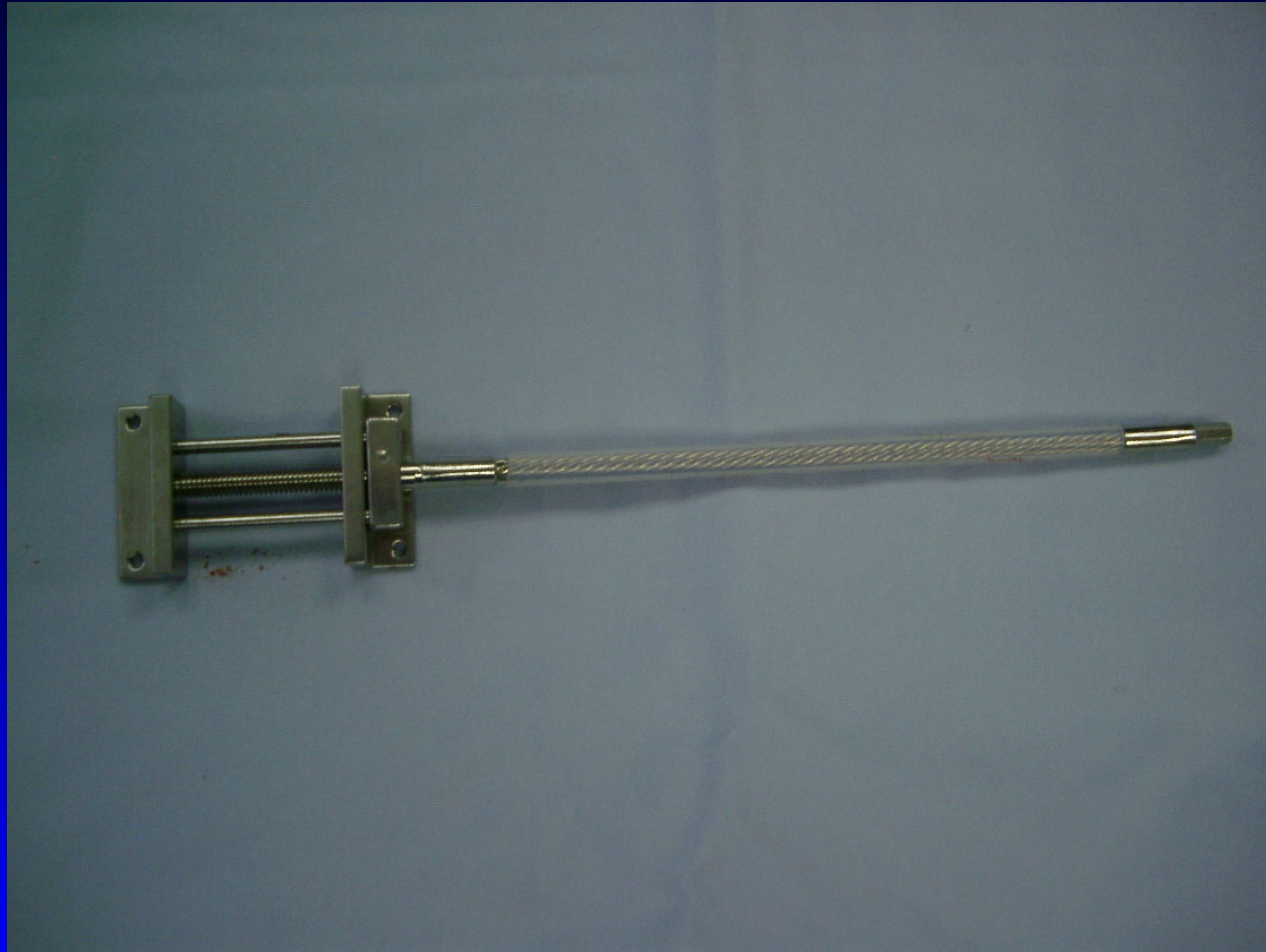


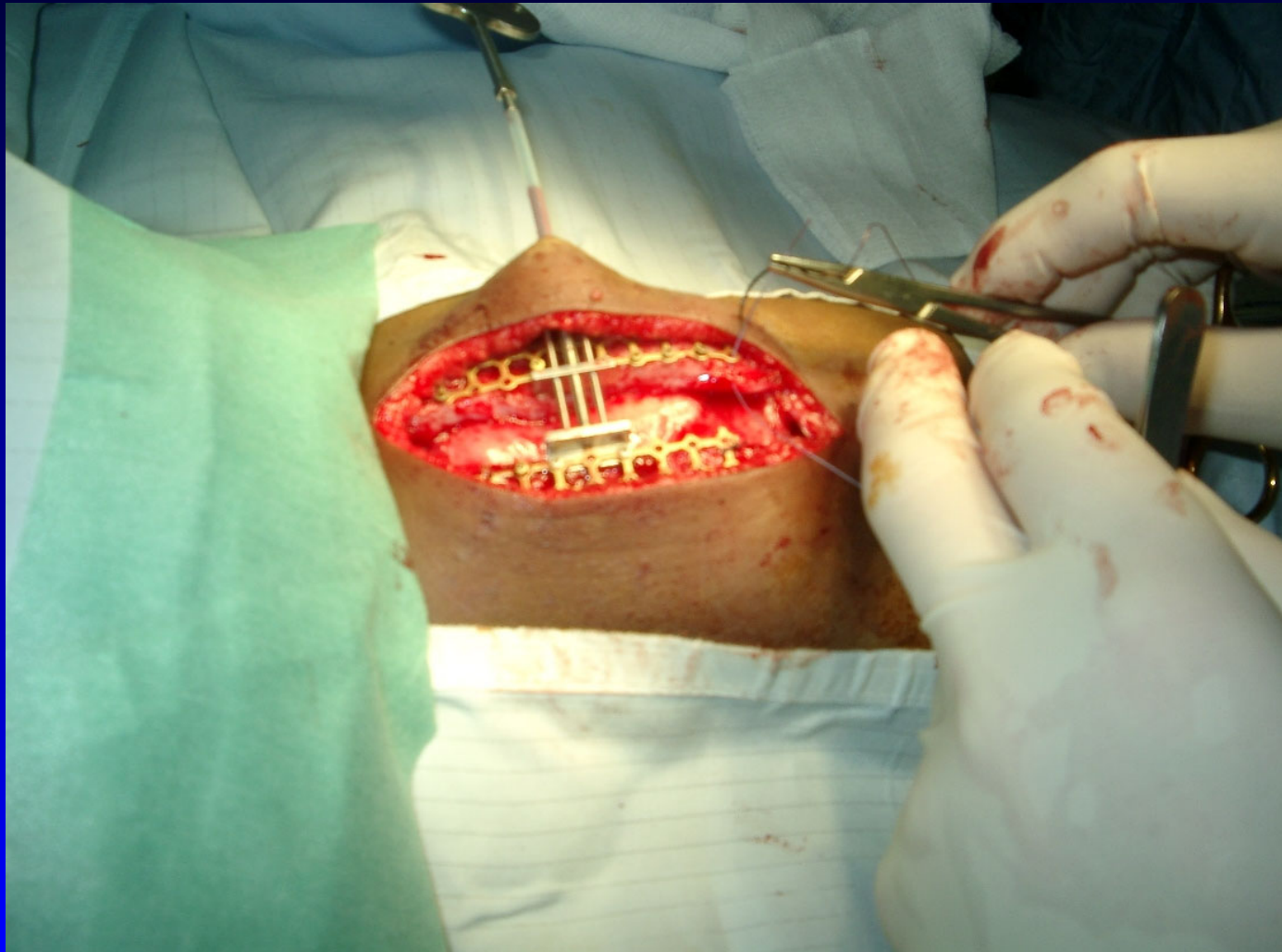






Leibinger maxillo-facial distractor



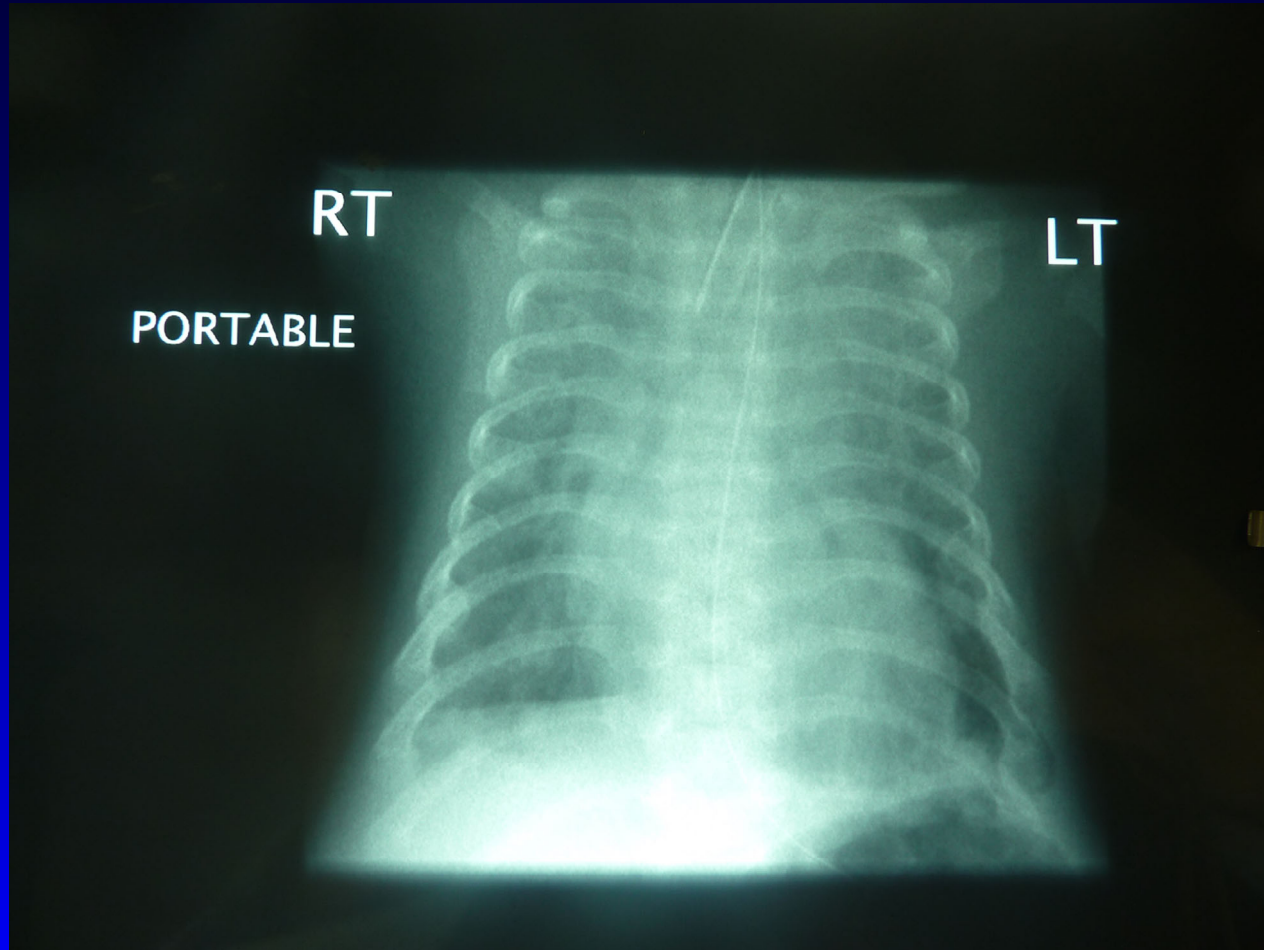


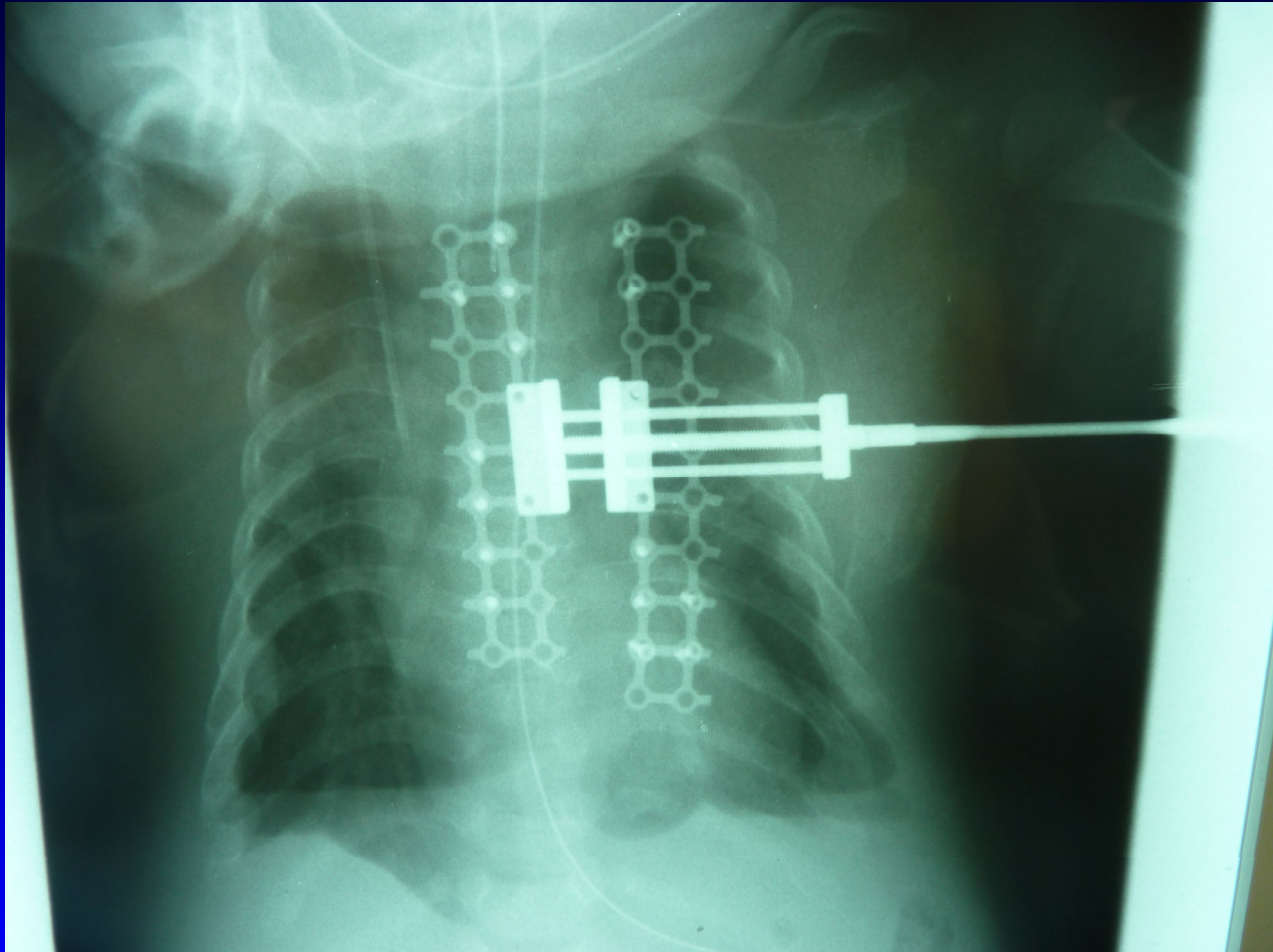


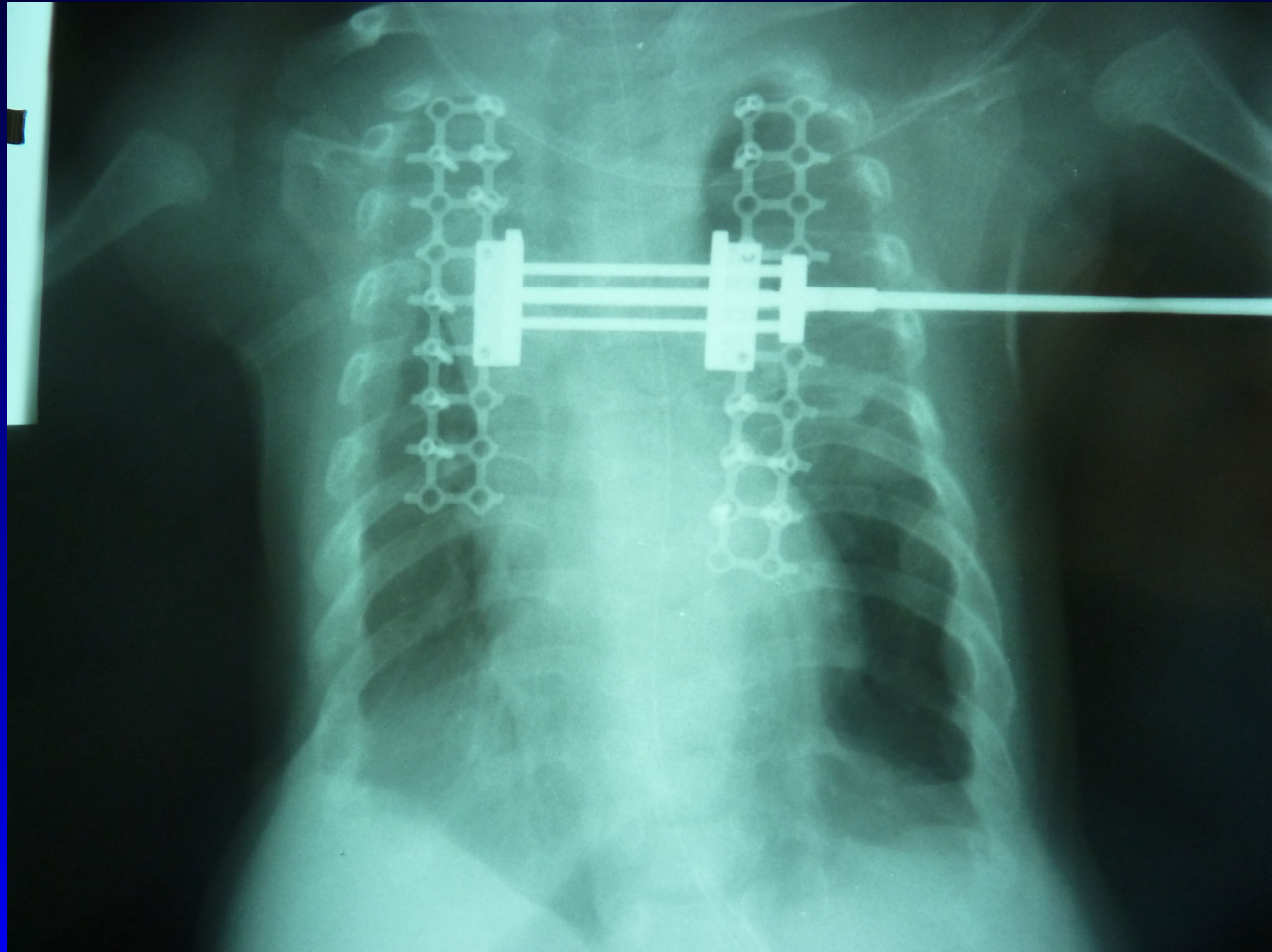


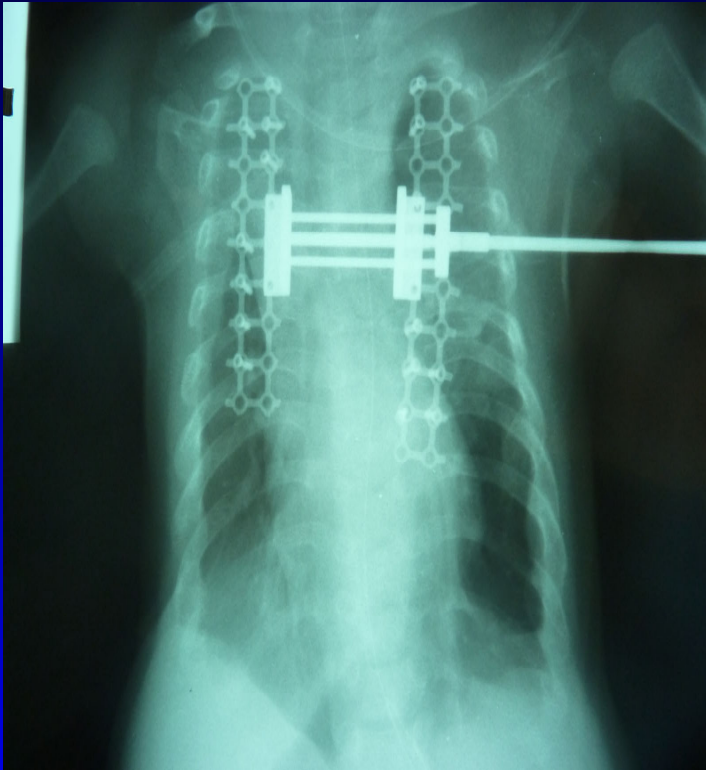


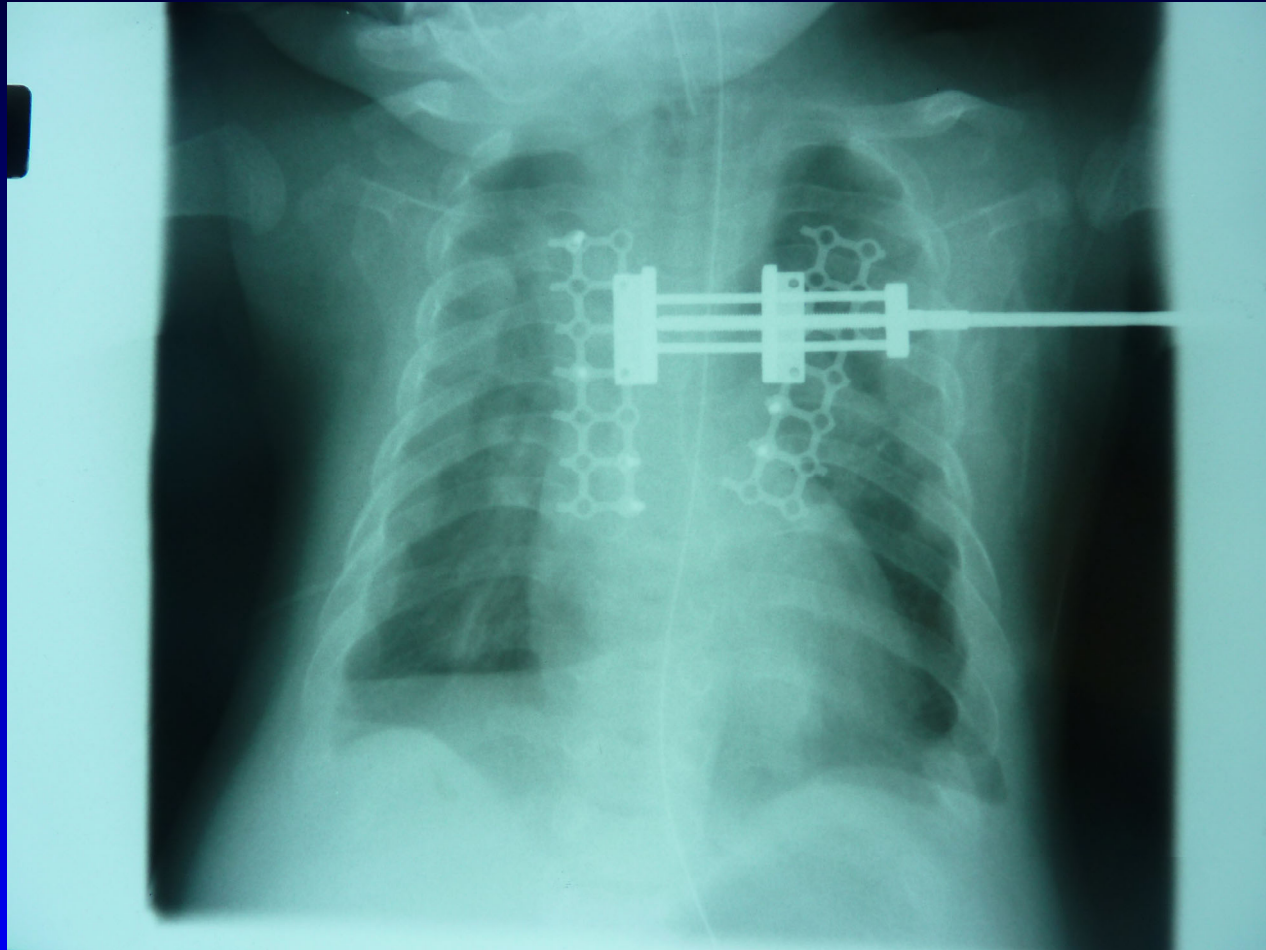
Pre sternotomy

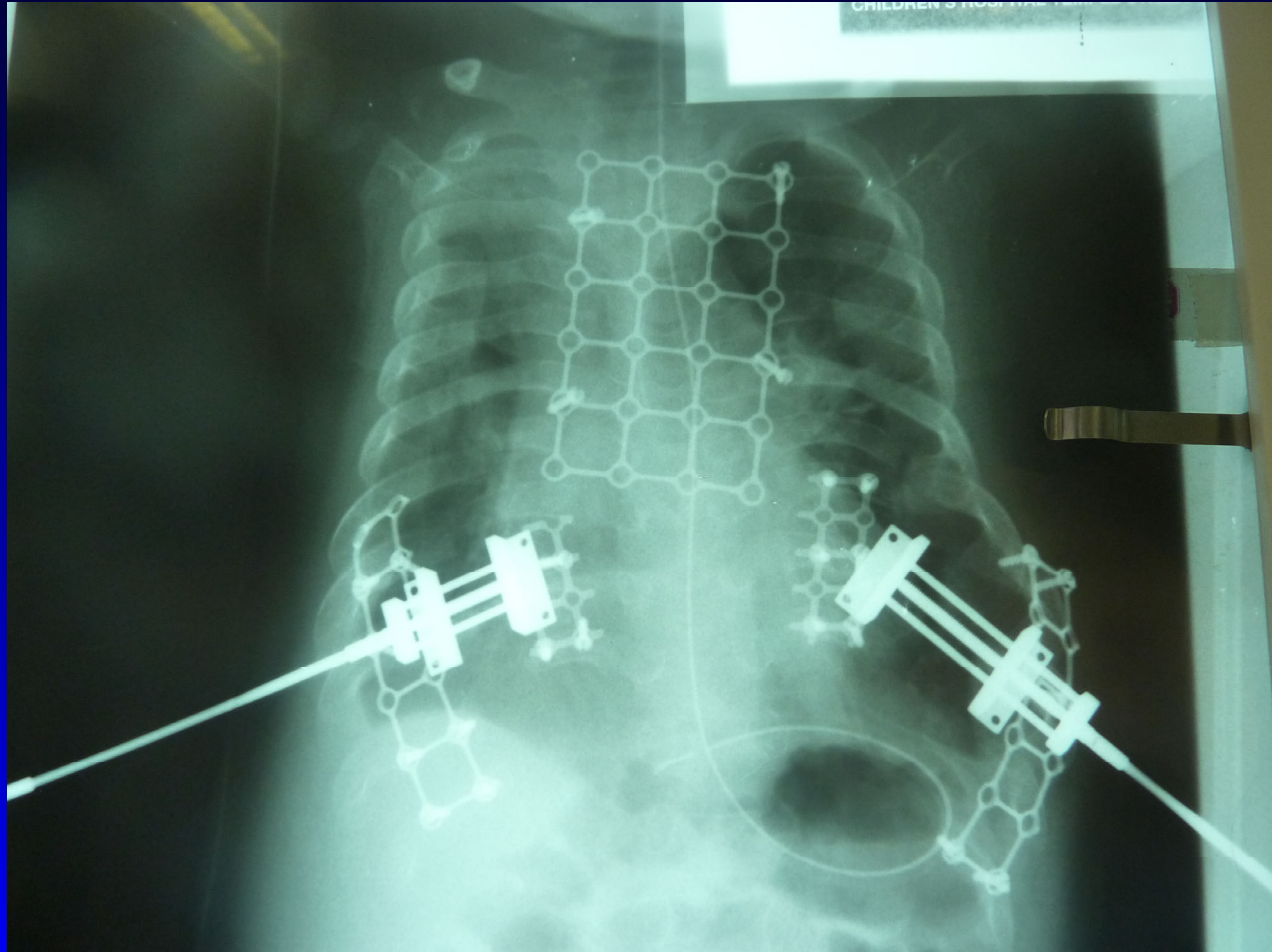


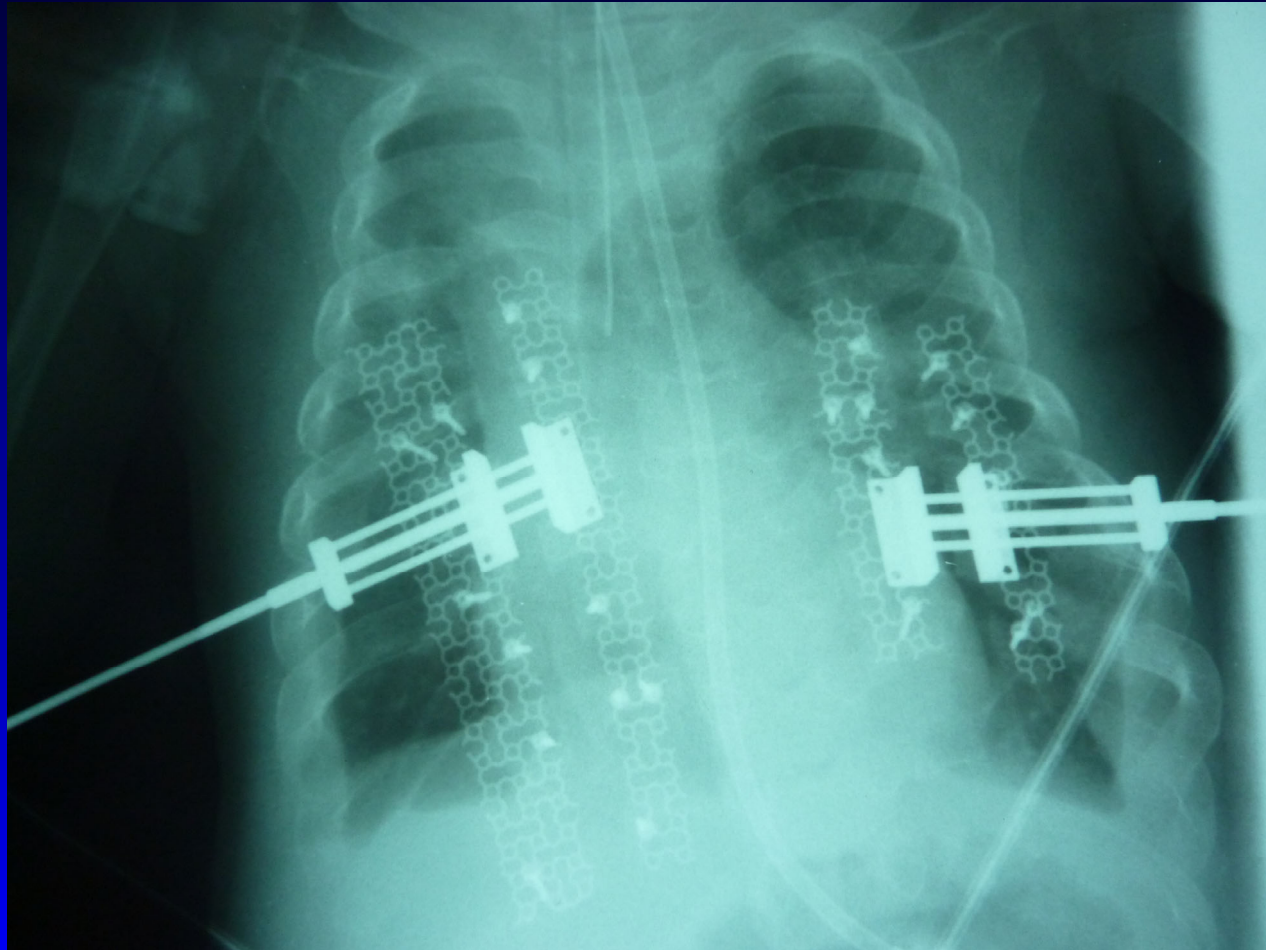


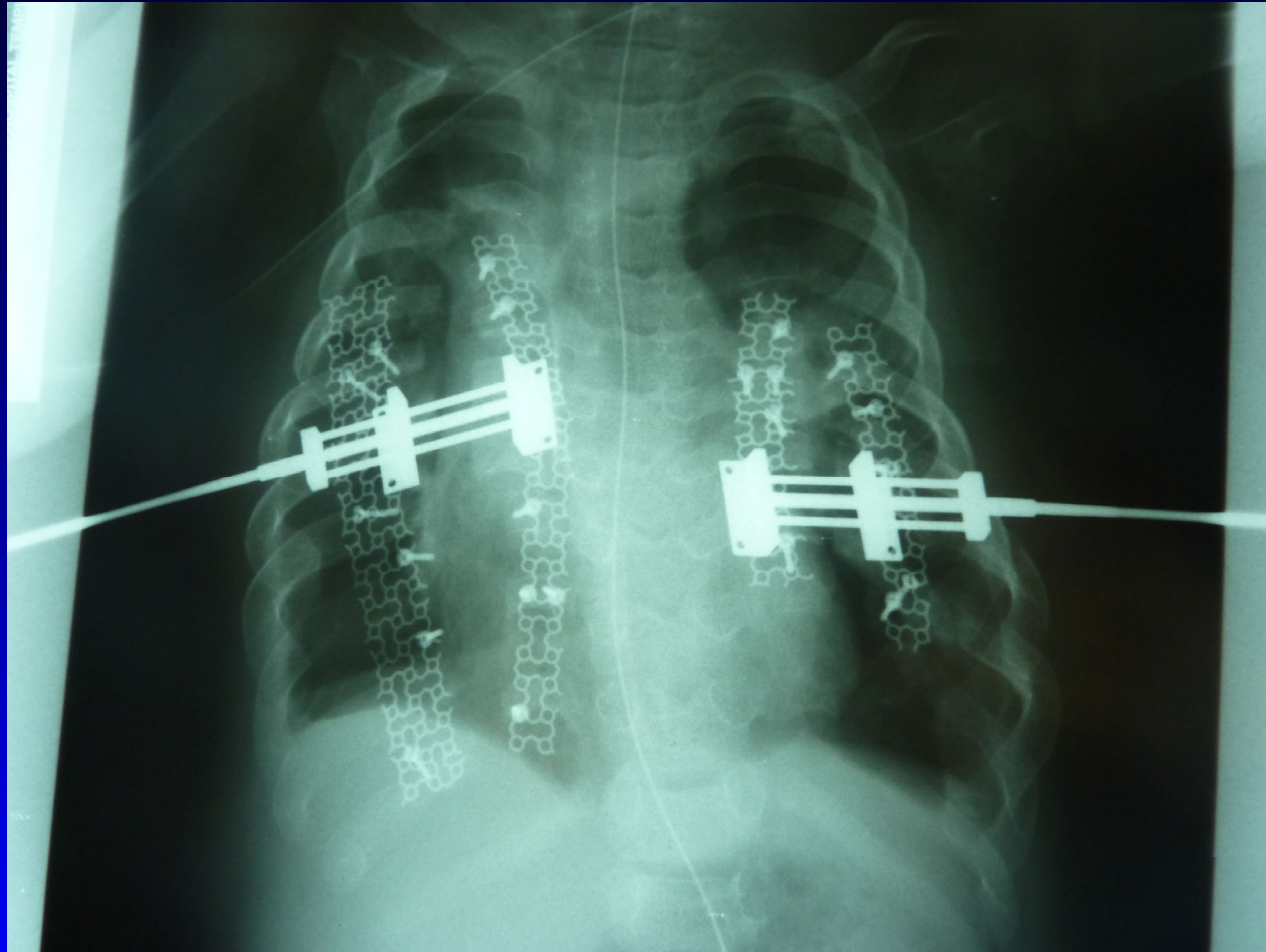


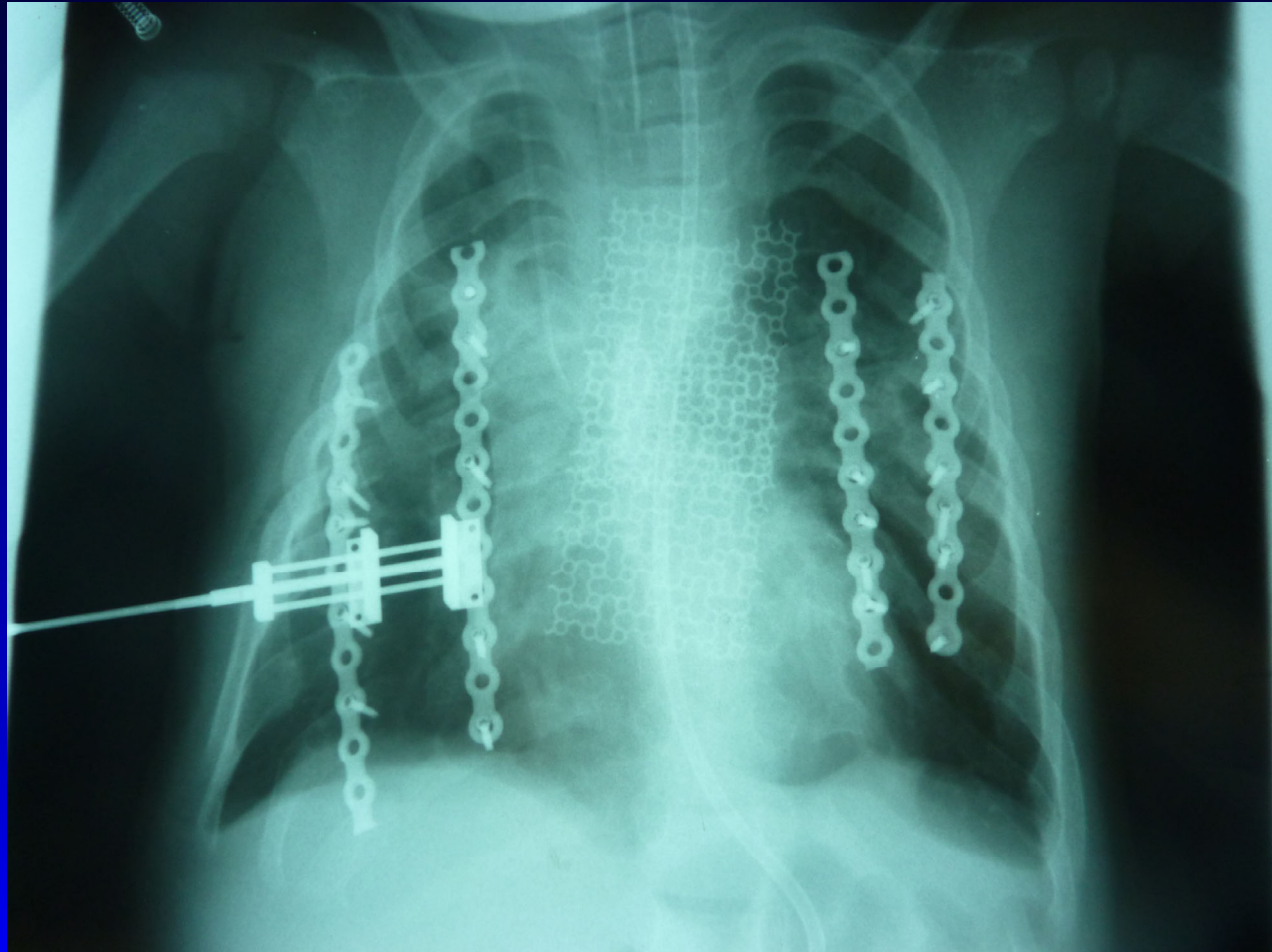


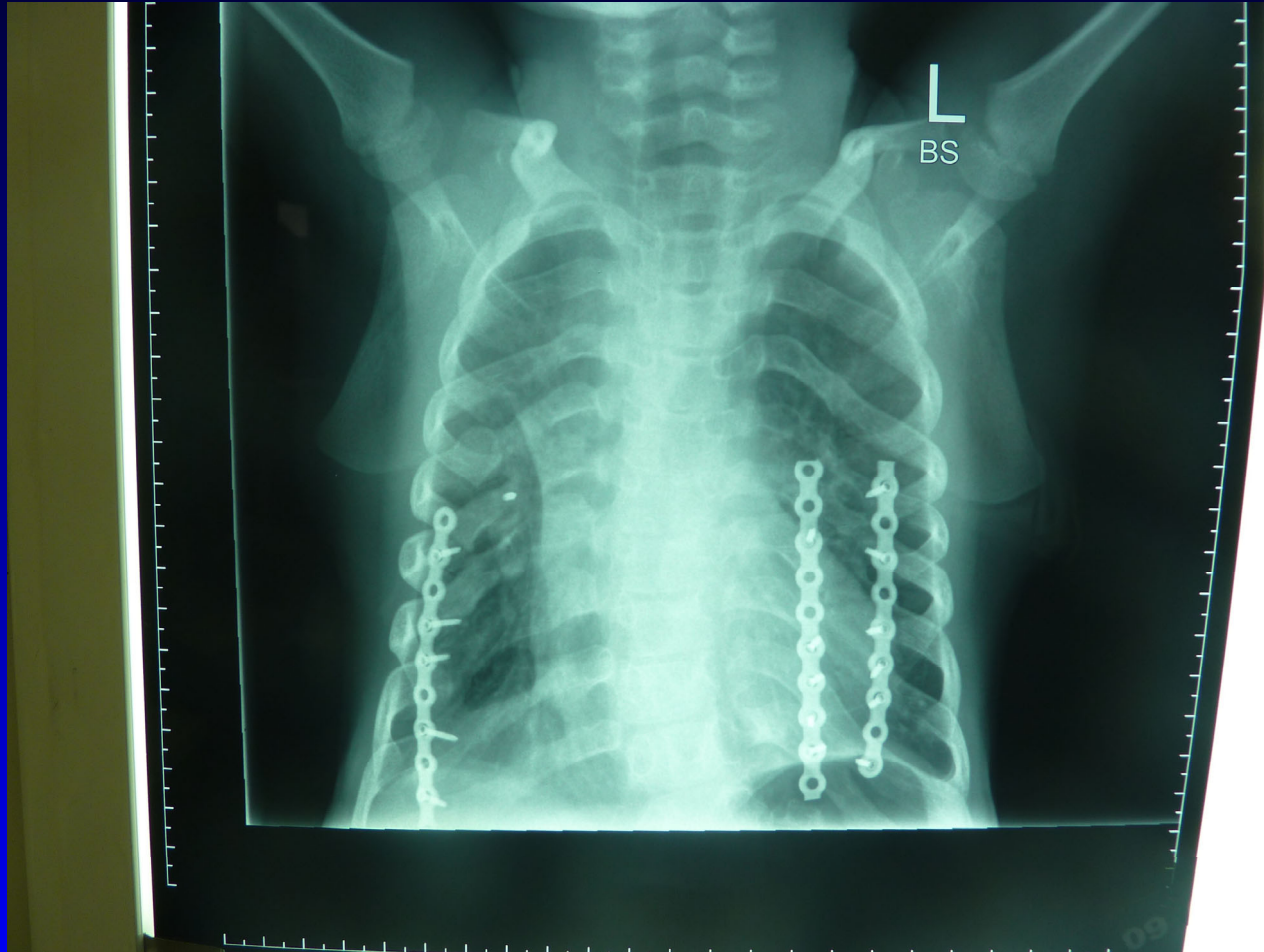


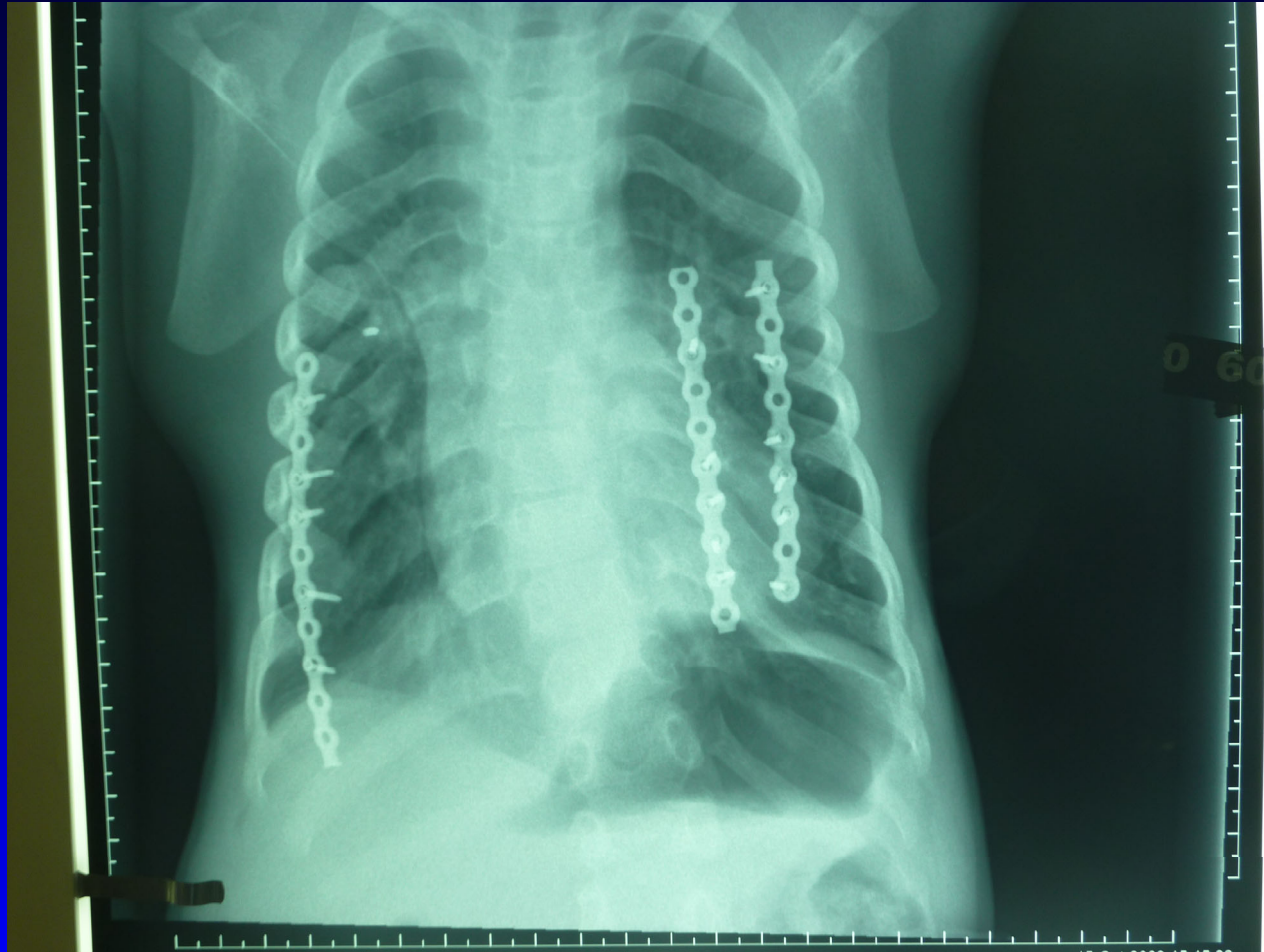






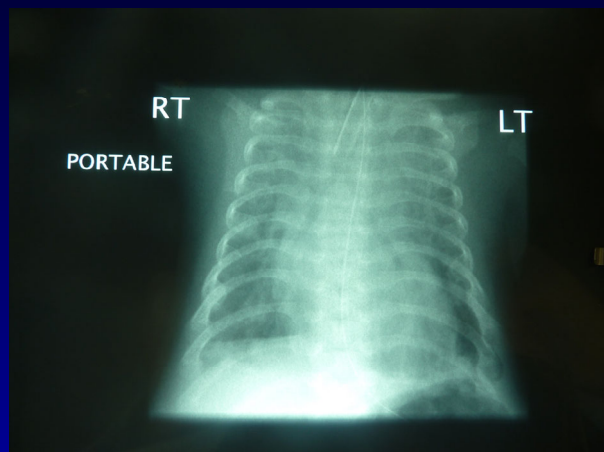




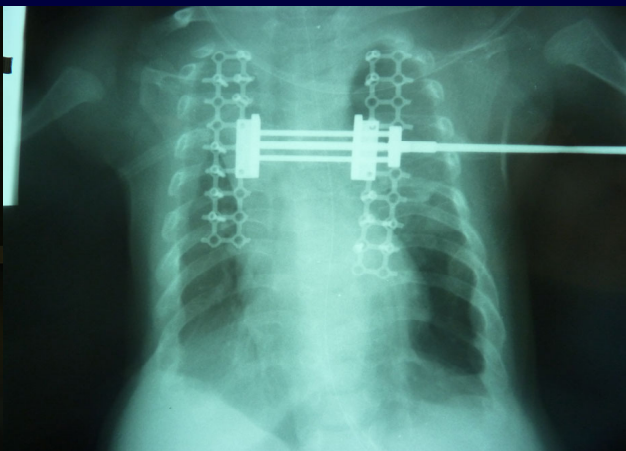


Dynamic thoracoplasty

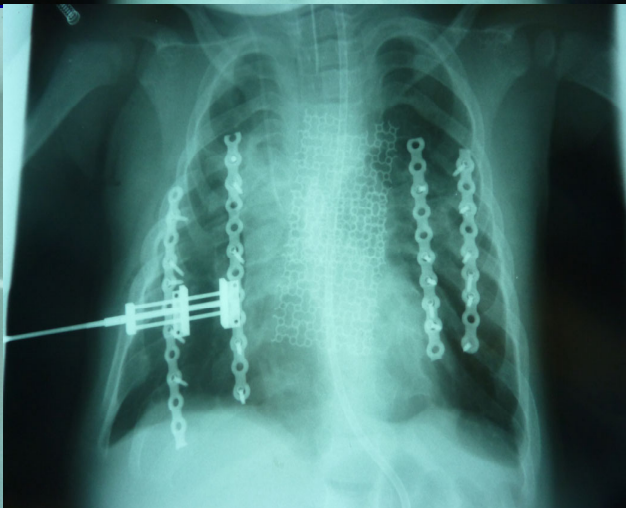
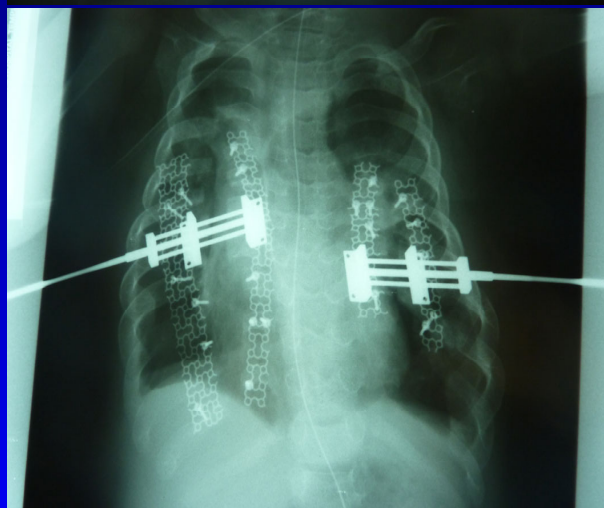
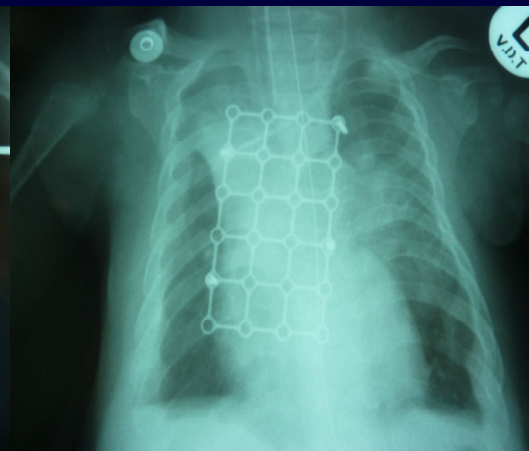
1/12



3/12



4/12



5/12

9/12

3.5 years

Dynamic Thoracoplasty

- Life saving in the neonate with severe Jeune syndrome
- Gradual distraction avoids skin /wound problems
- x10 increase in alveoli in the first 2 years of life
- Gradual distraction “grows the lung” not just the chest wall
- ? Applicable also in severe neonatal spinal deformity

Thank you

