

Professor Damian McCormack

Paediatric Orthopaedic Surgeon

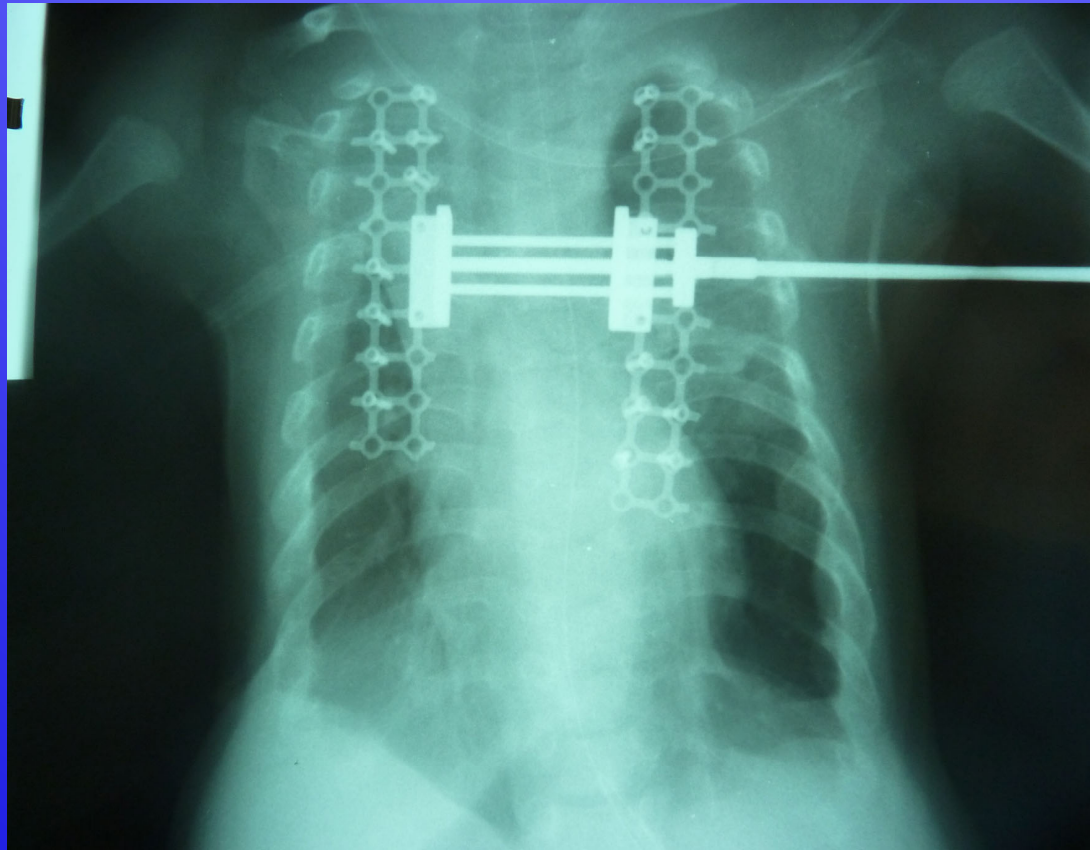
I have No commercial confessions

Dr Ali Alekri

I dedicate this talk to my
good friend and former
Fellow currently on hunger
strike in Jaw prison in
Bahrain



Sternal Spread and Rib procedures for Juene and like dysplasias



Prof Damian Mc Cormack
Dublin

Juene Syndrome

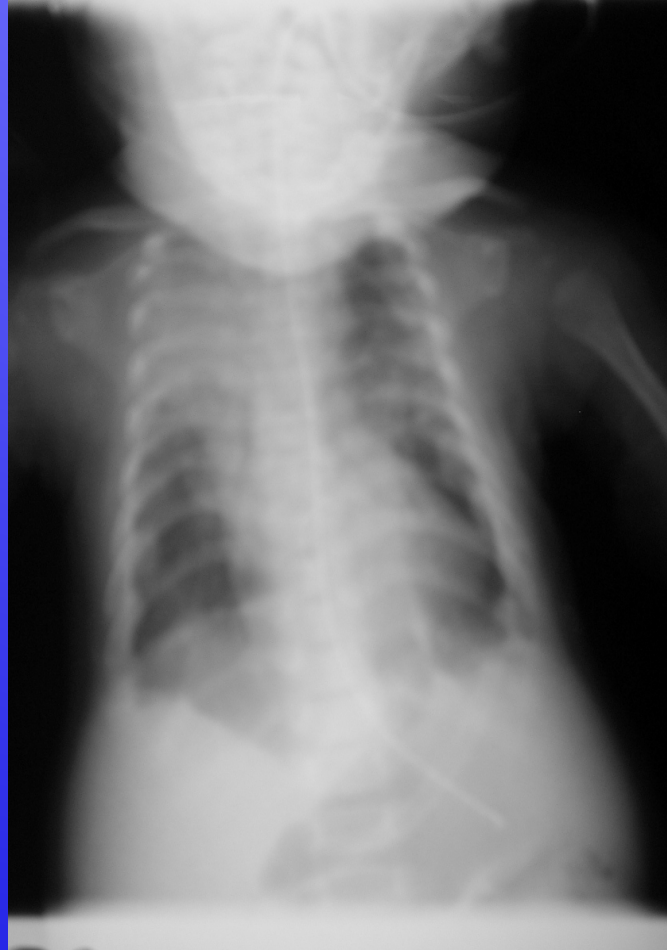
- Familial Asphyxiating Thoracic Dystrophy
- Juene 1955
- Rare $1 / 10^5$ live births
- Autosomal recessive acromelic dwarfism
- Variable severity reflects genetic heterogeneity
- Restrictive lung disease
- Renal / pancreatic disease
- Neonatal Juene Syndrome is fatal in 70 %

Juene syndrome

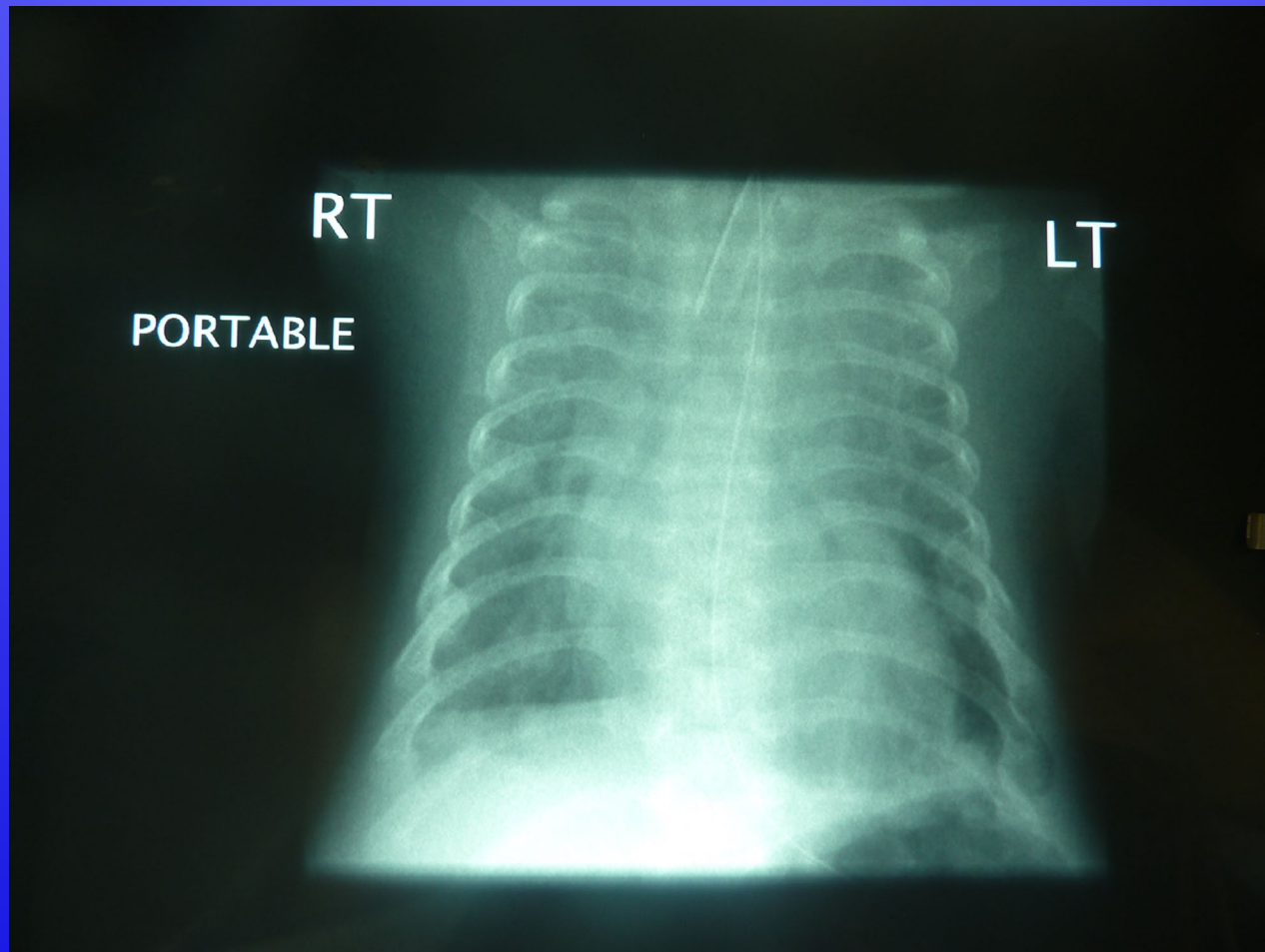
Locus at chromosome
15q13
Locus at 3q24-3q26

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

Nat Genet Jun 2007

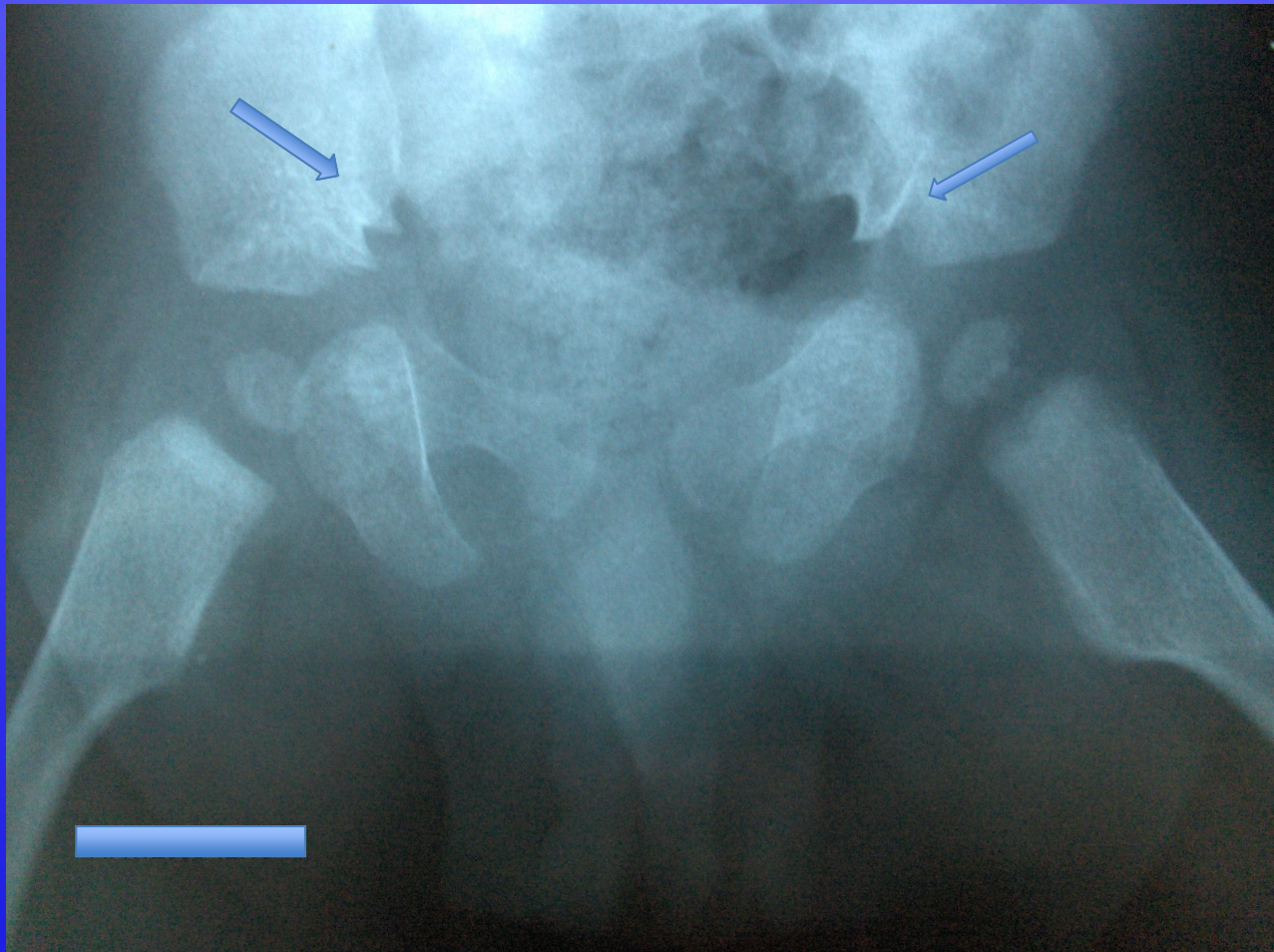


Juene syndrome

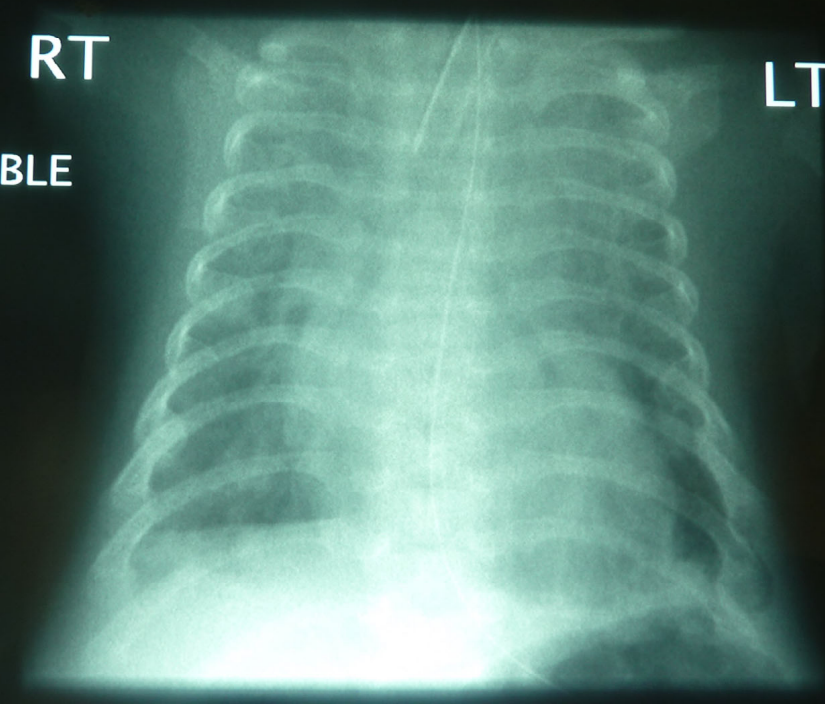


Juene syndrome

sciatic notch bone spikes

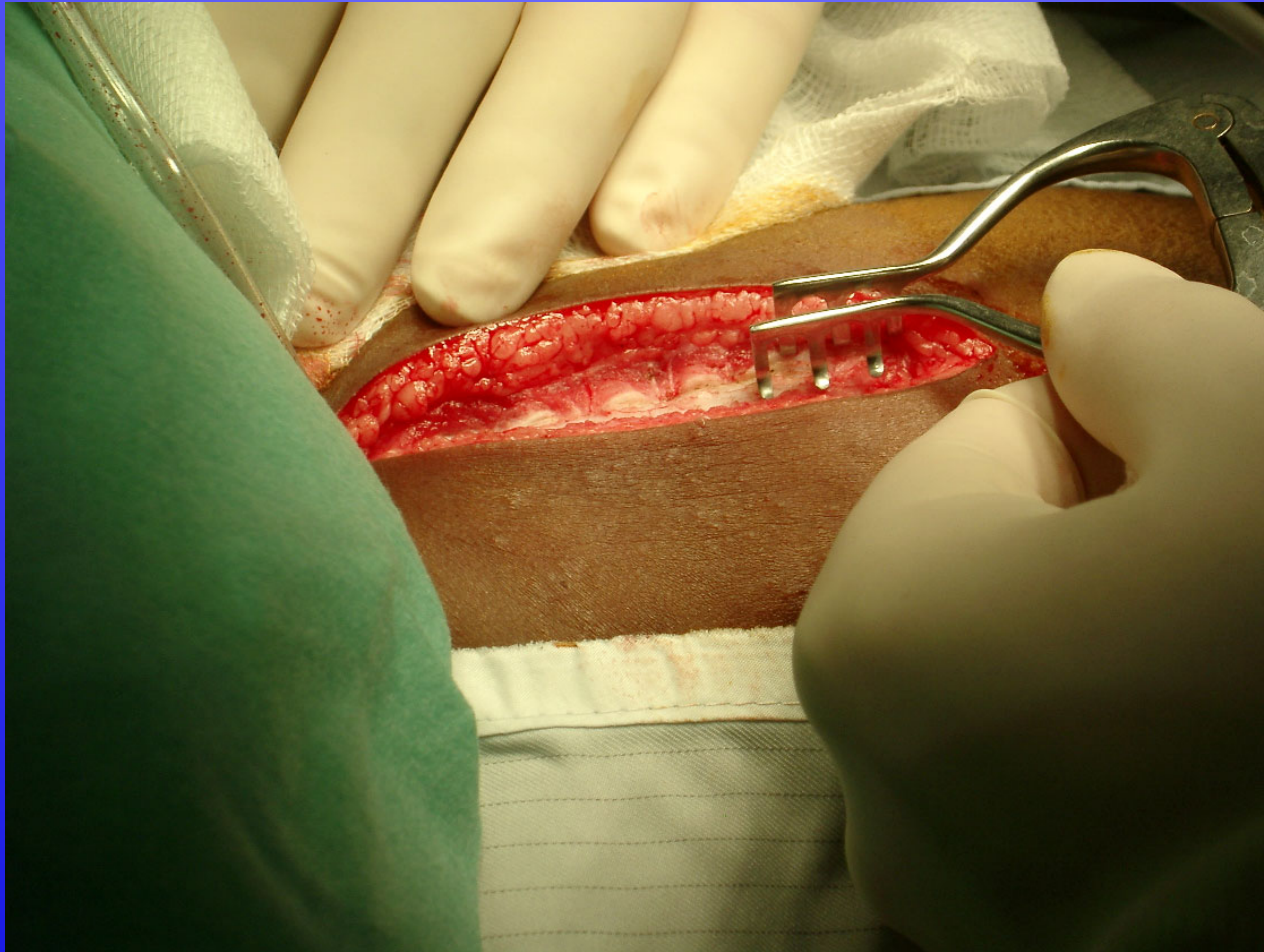


RT
PORTABLE
LT

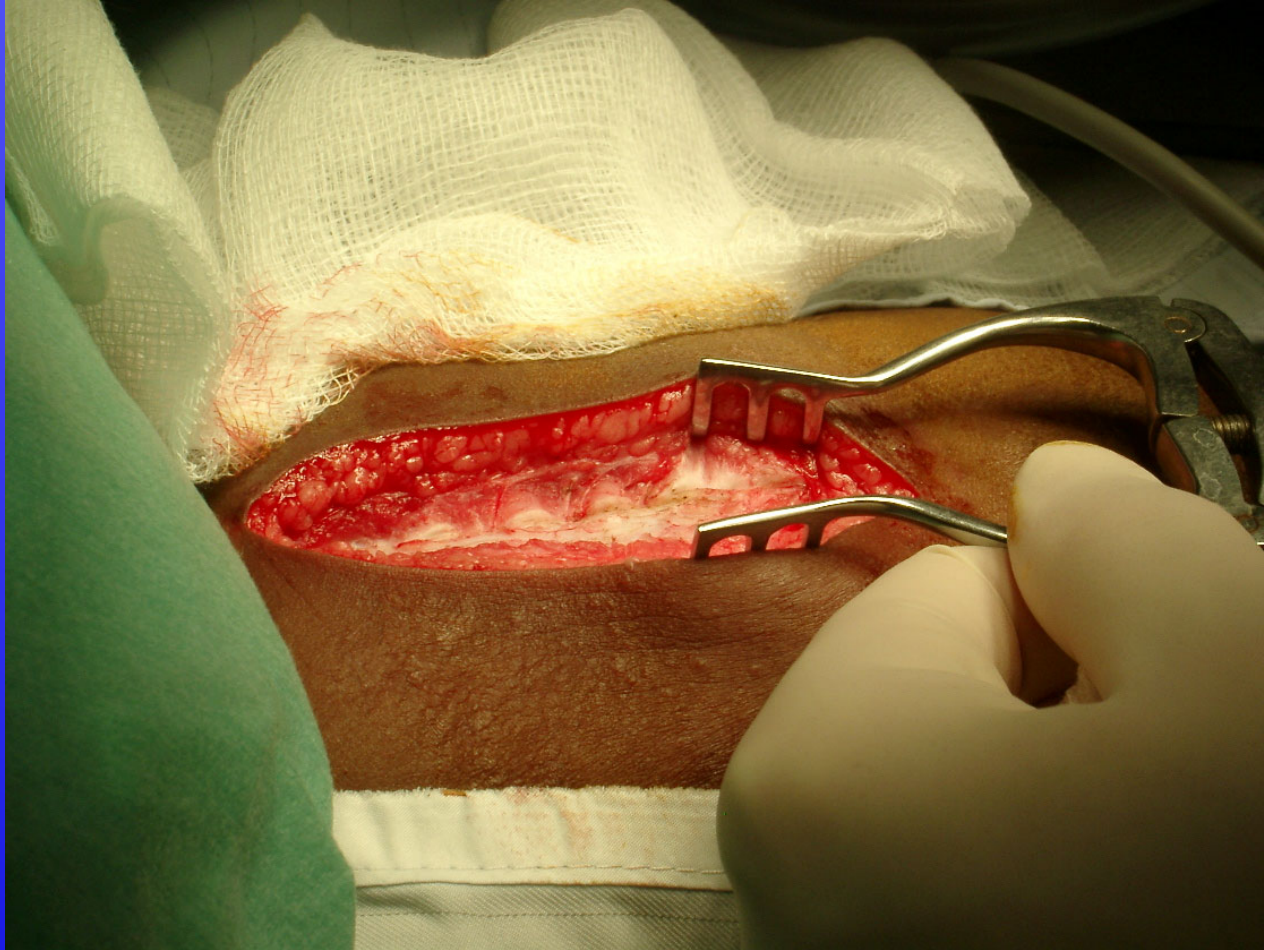




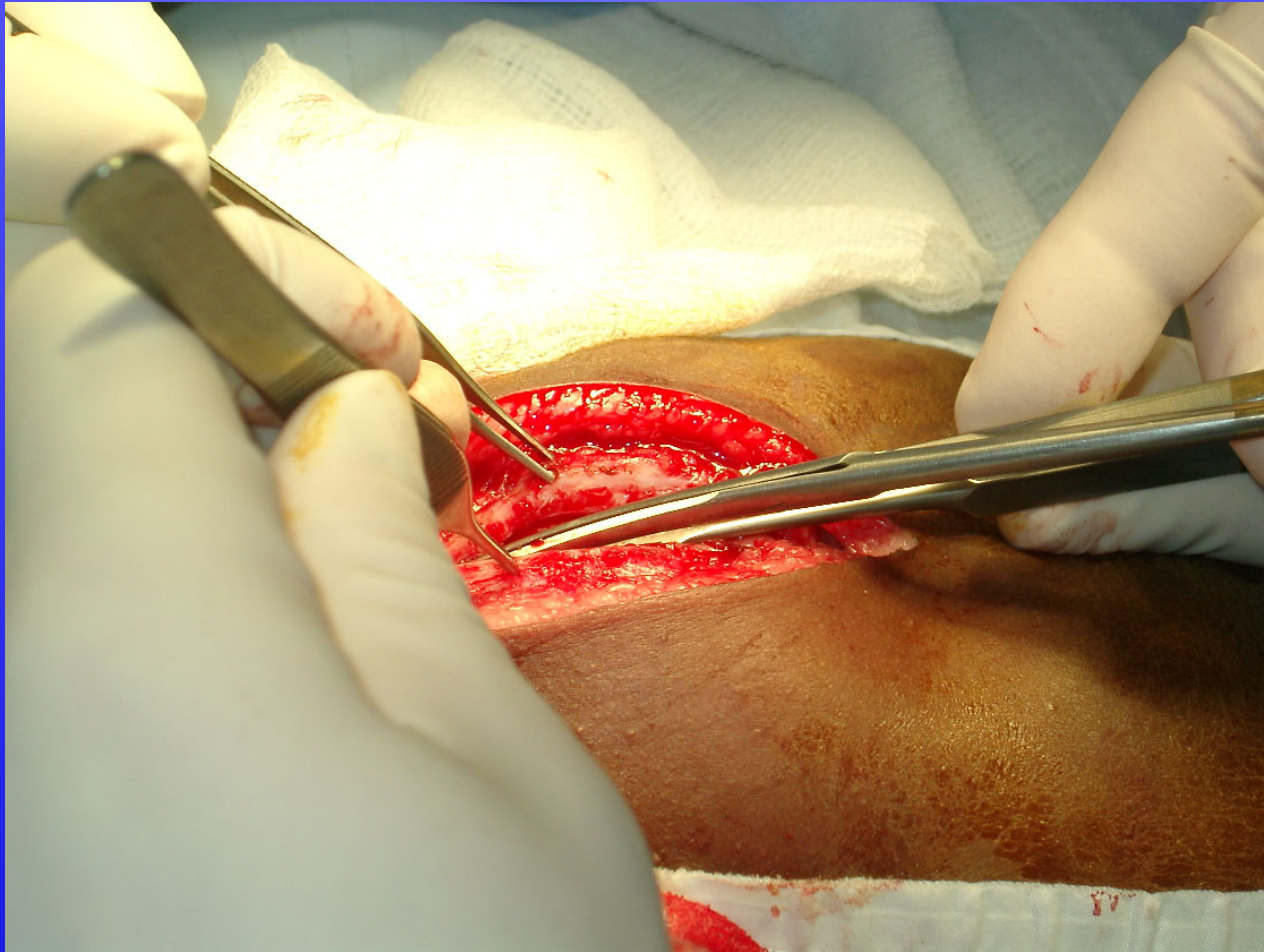
sternotomy

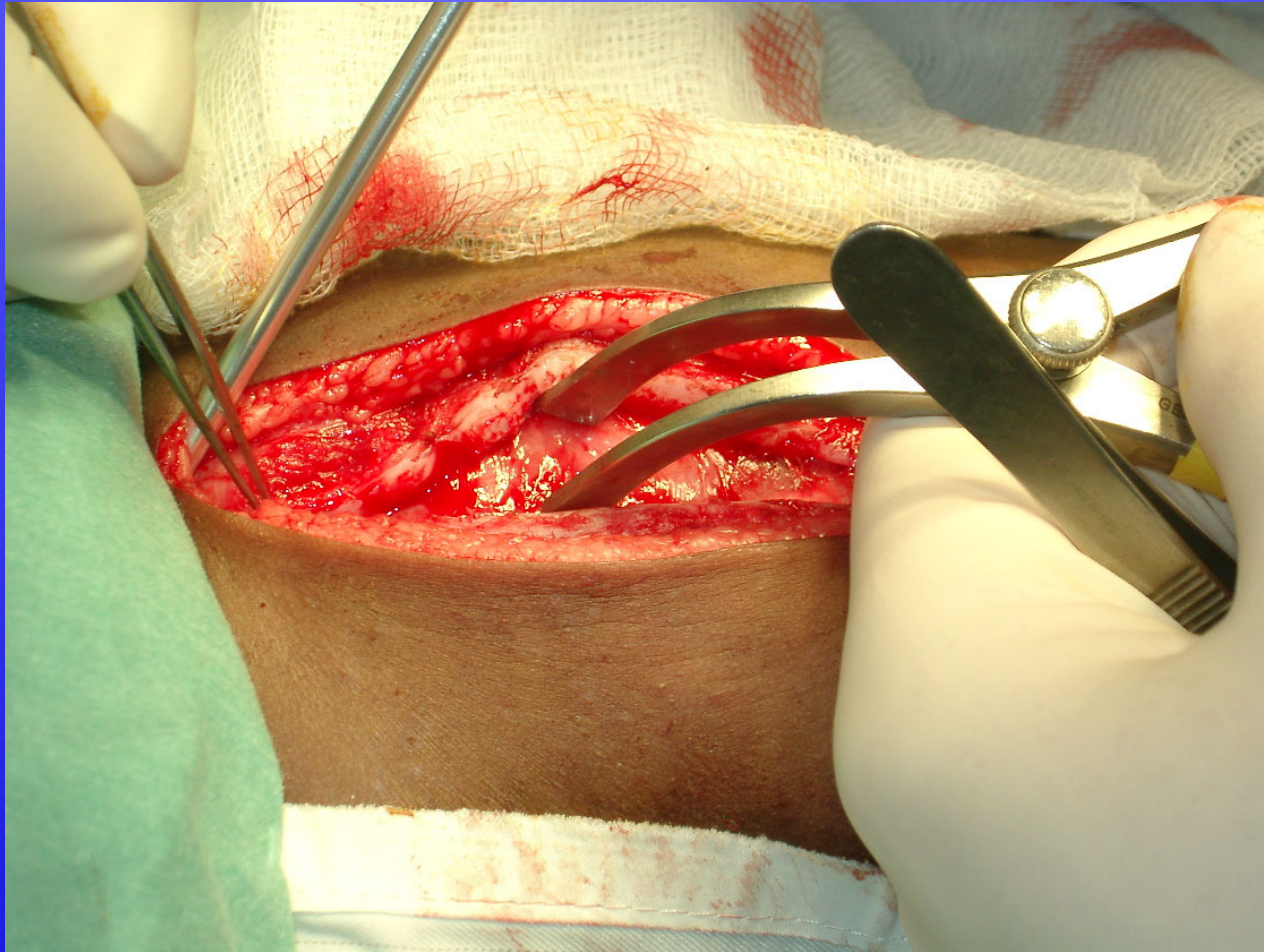


sternotomy

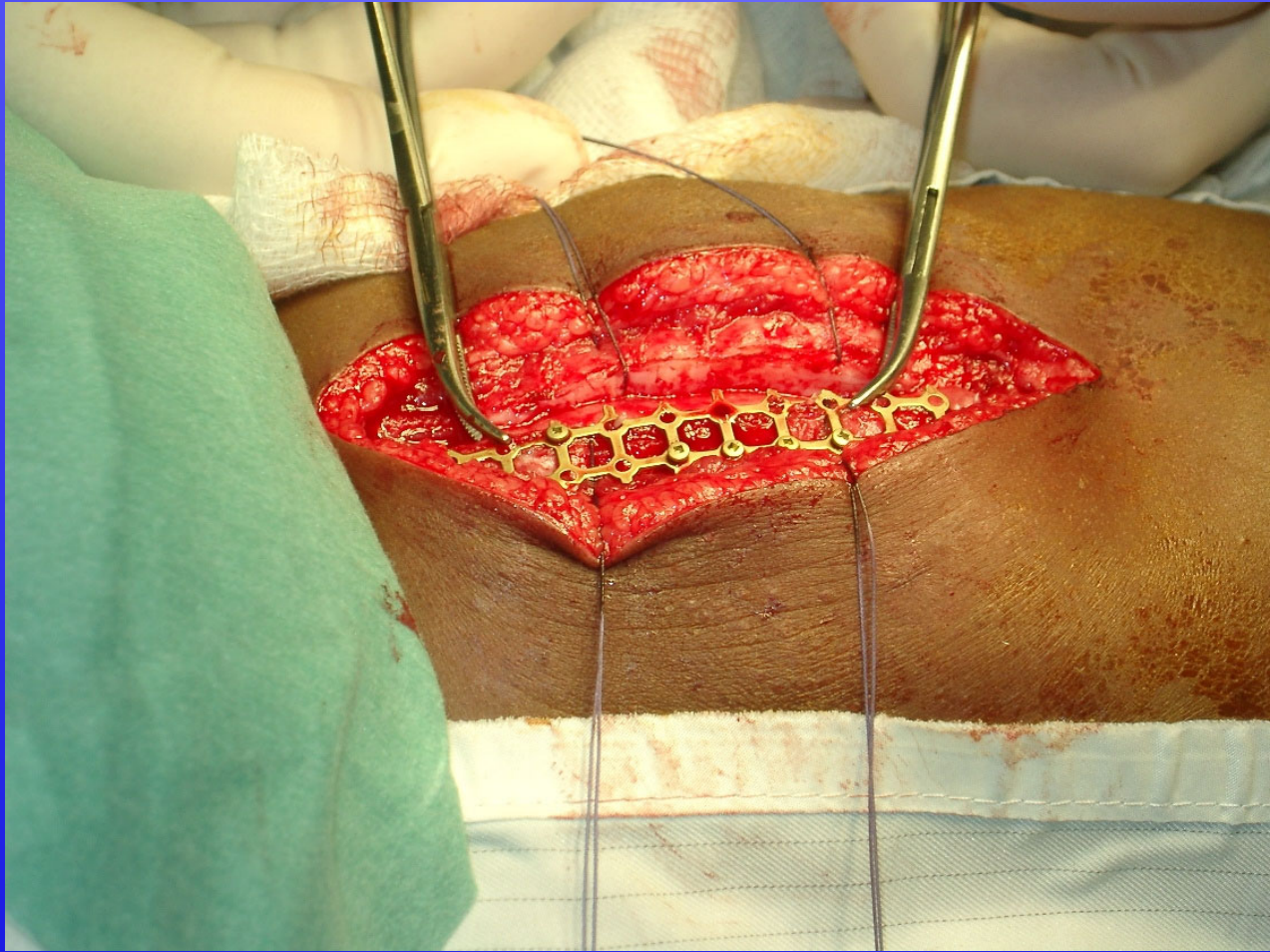


sternotomy

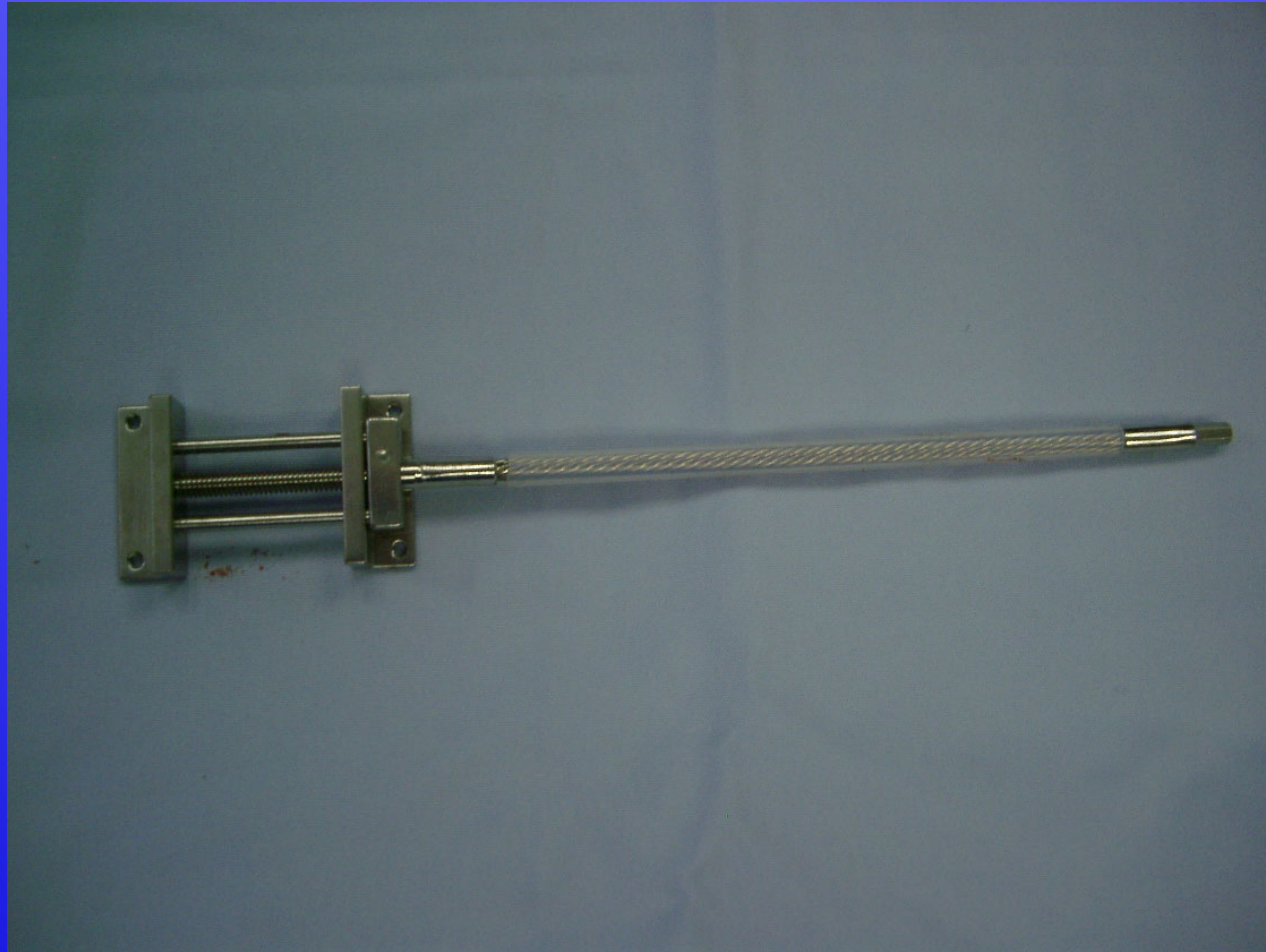




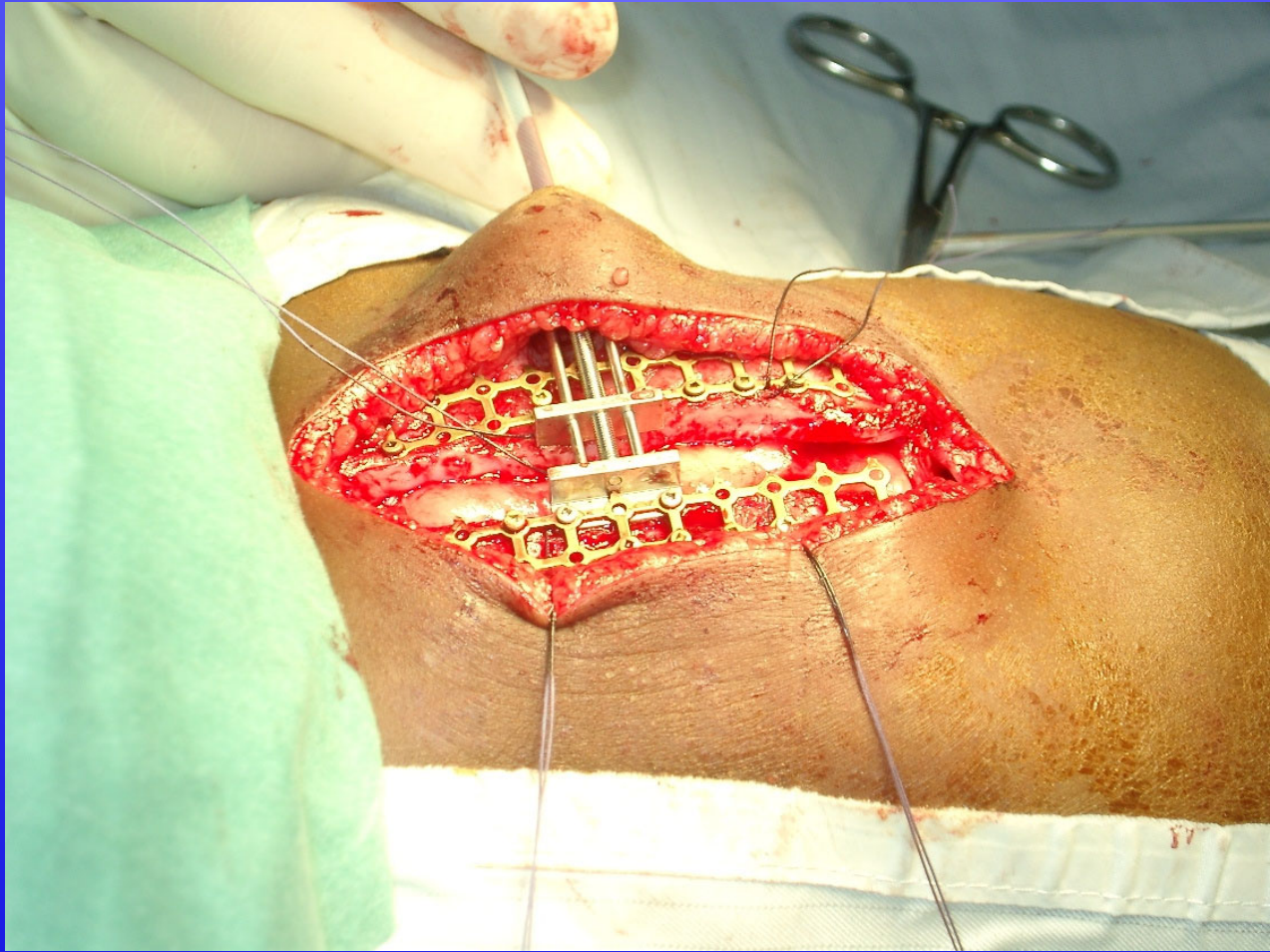




Leibinger maxillo-facial distractor



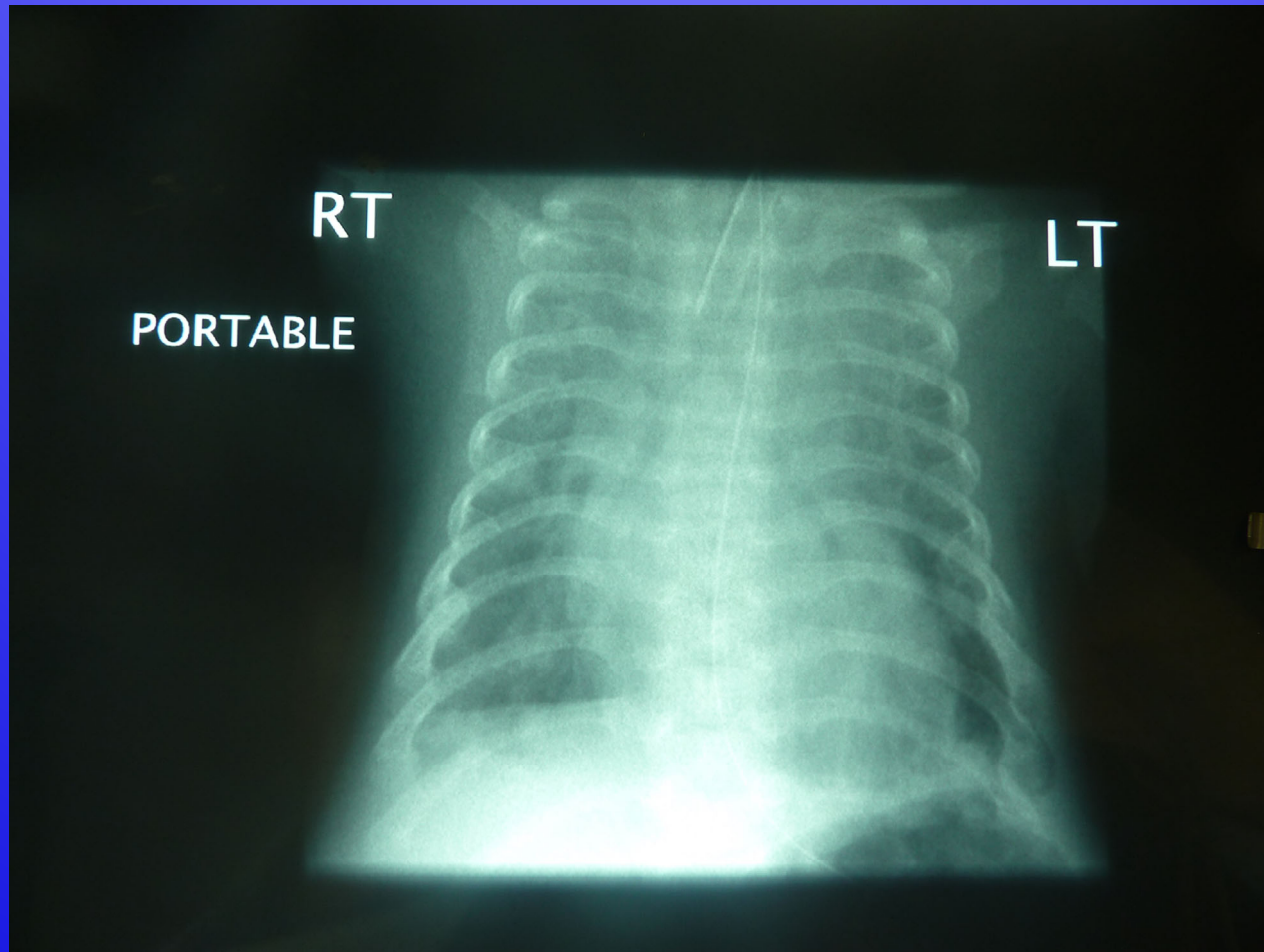




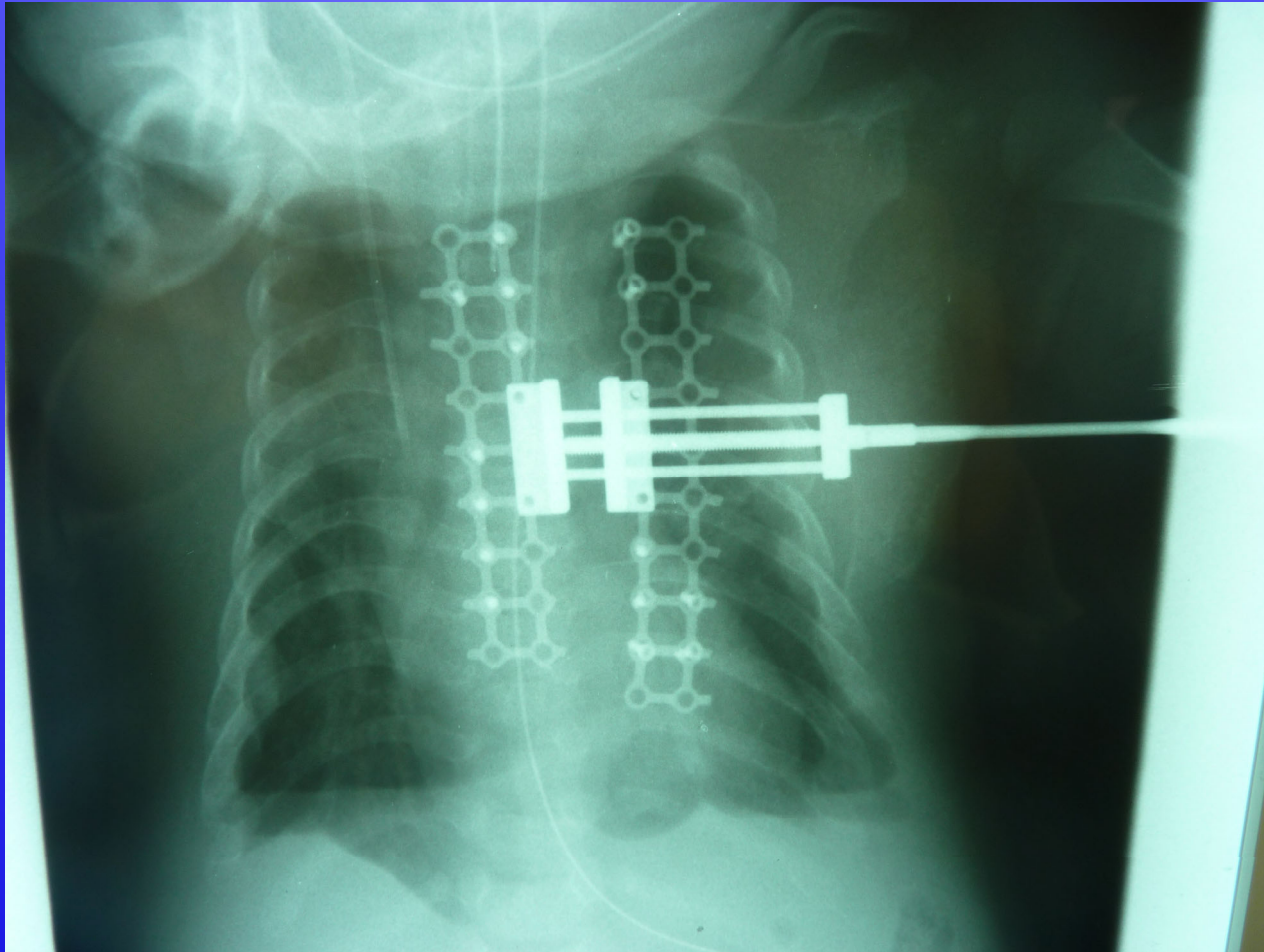




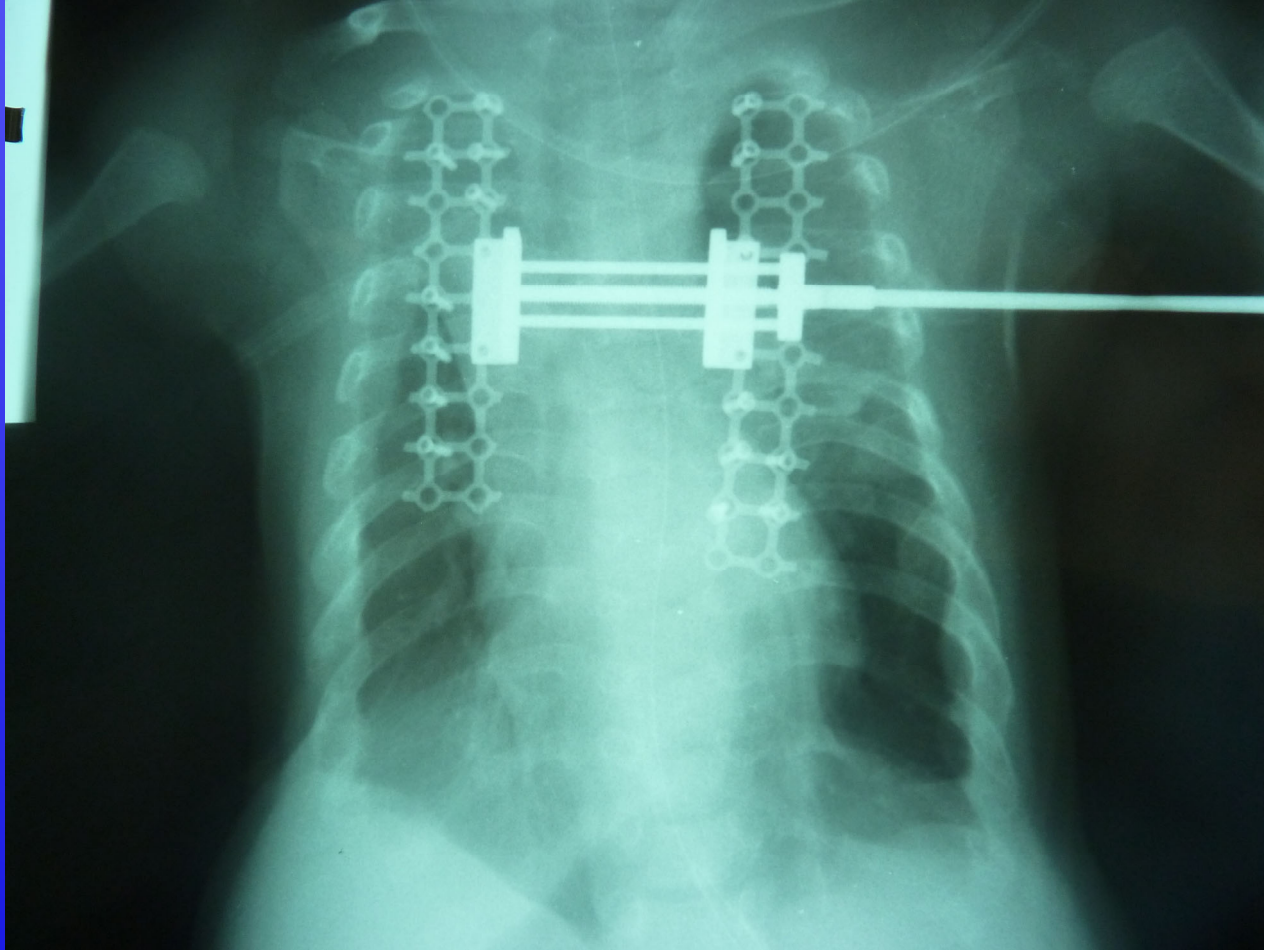
Pre sternotomy

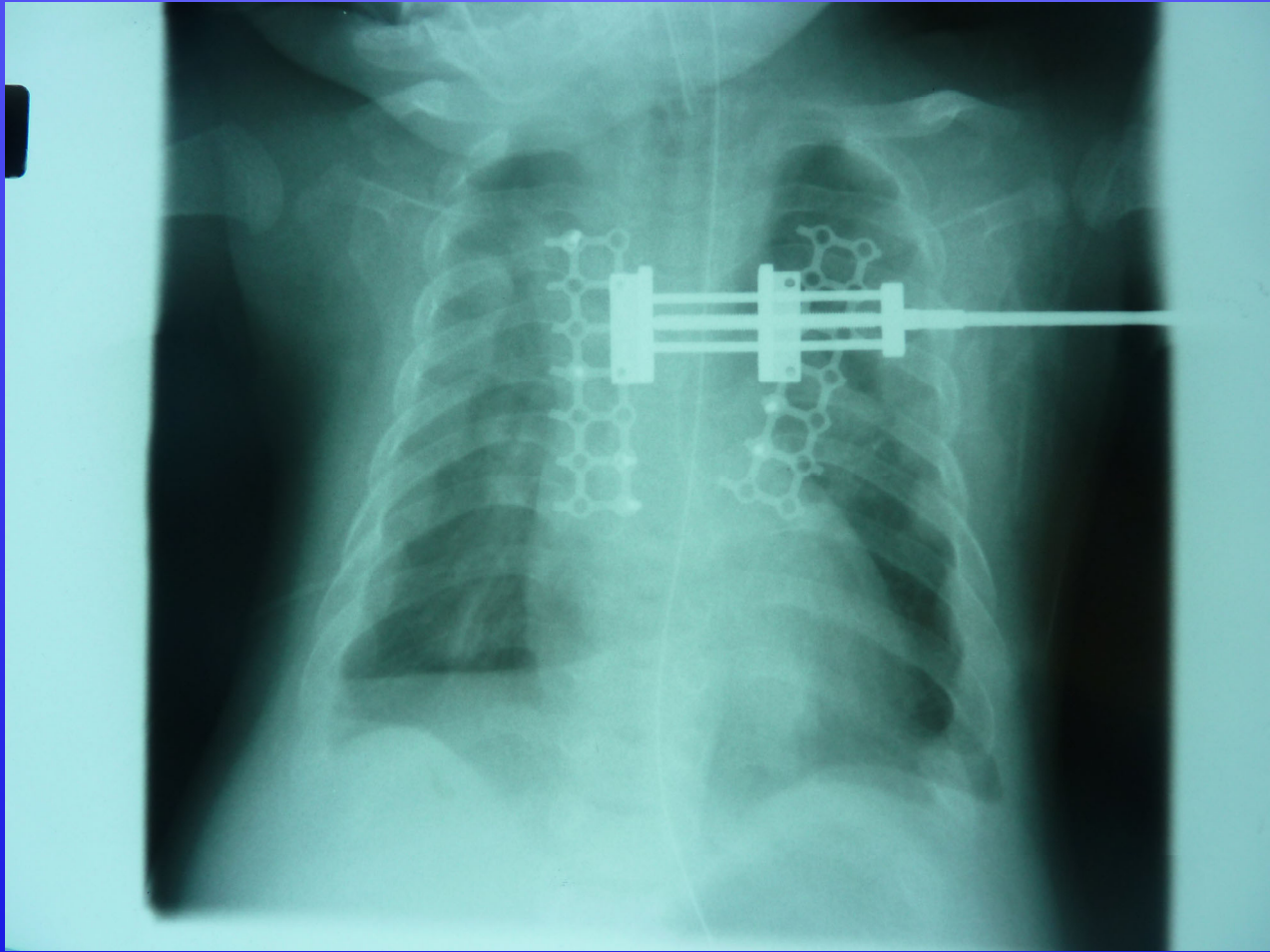


Post sternotomy

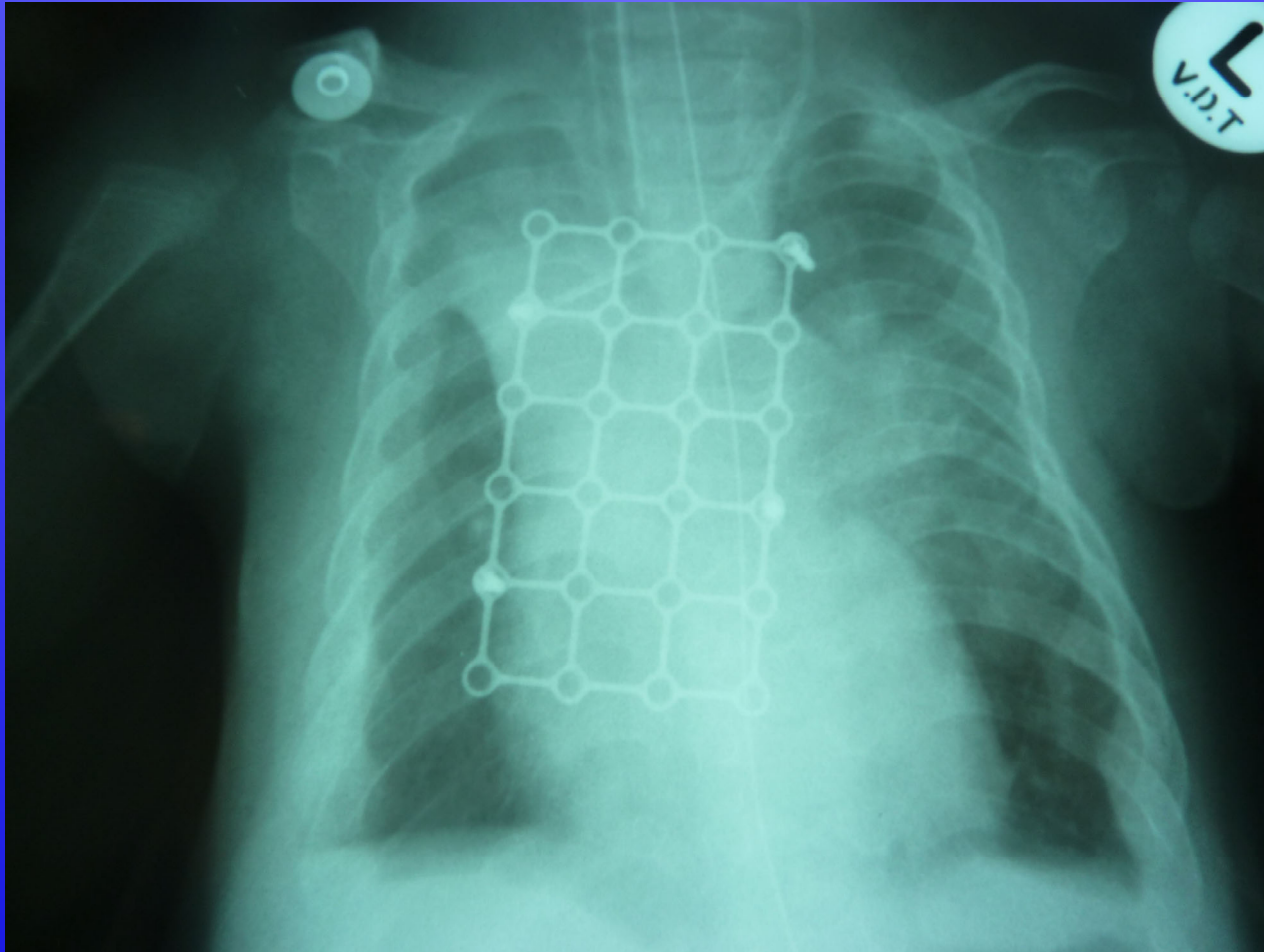


Sternal distraction

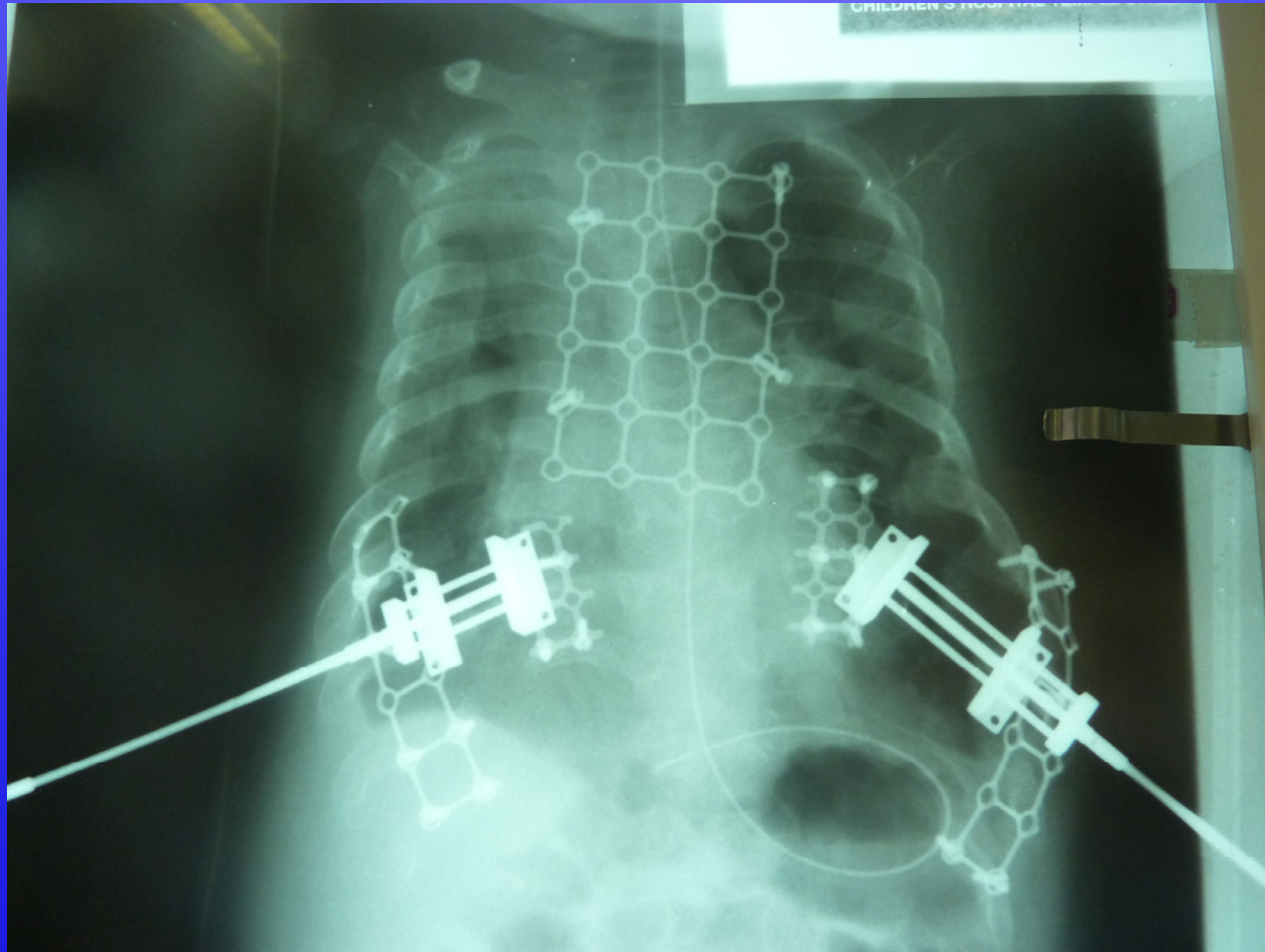




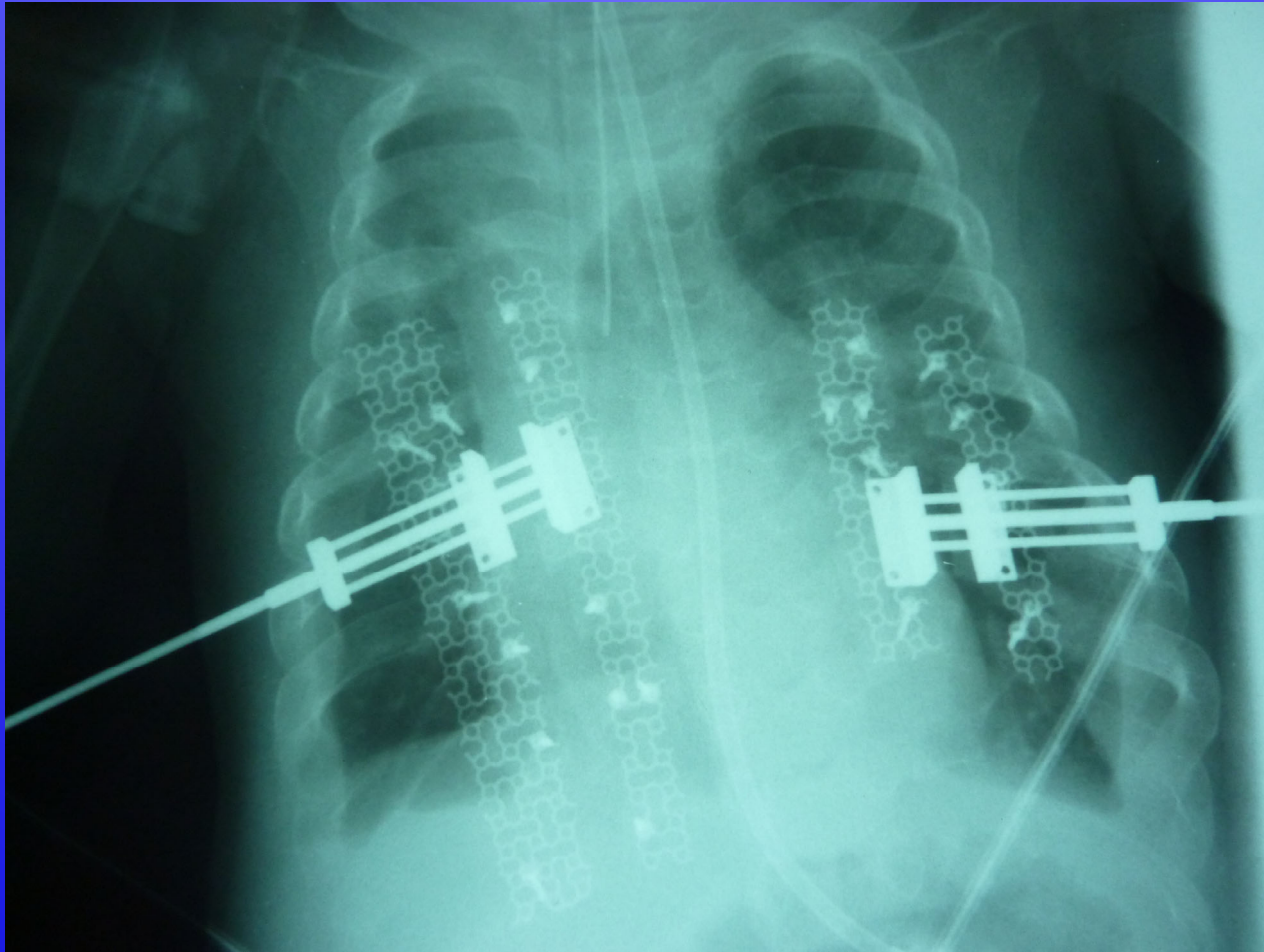
Static sternal distraction



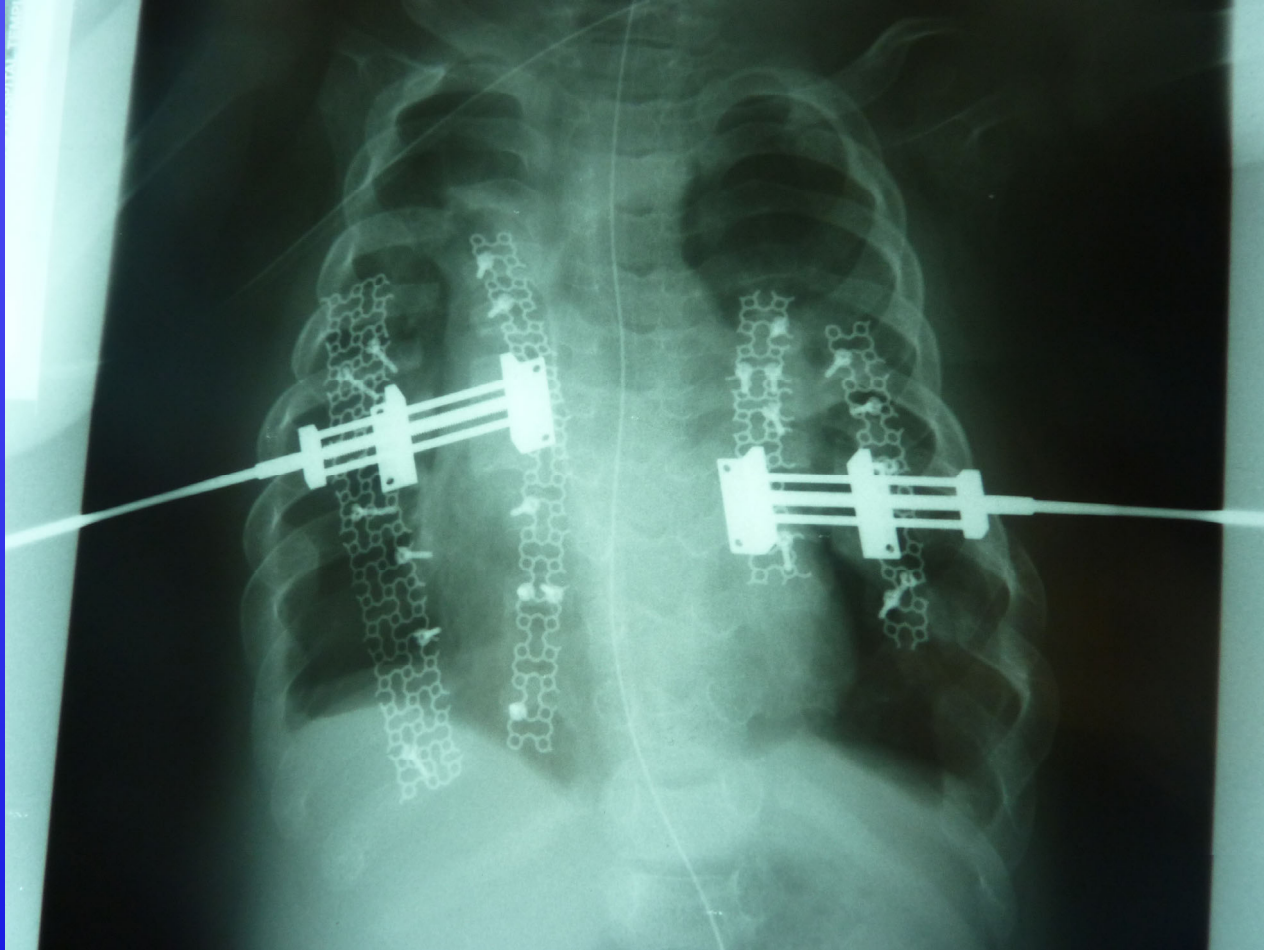
Rib distraction -osteogenesis



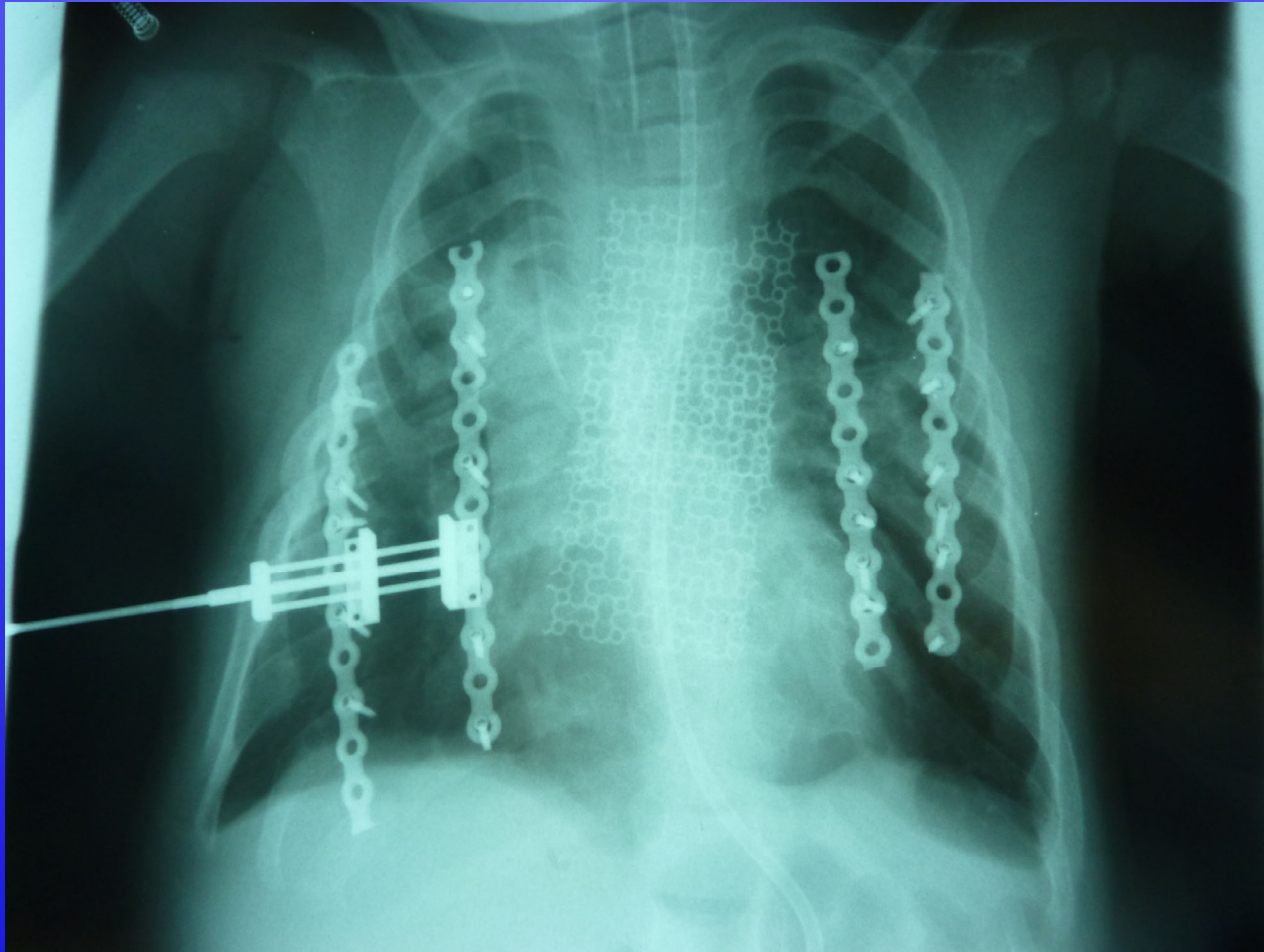
Rib distraction - osteogenesis

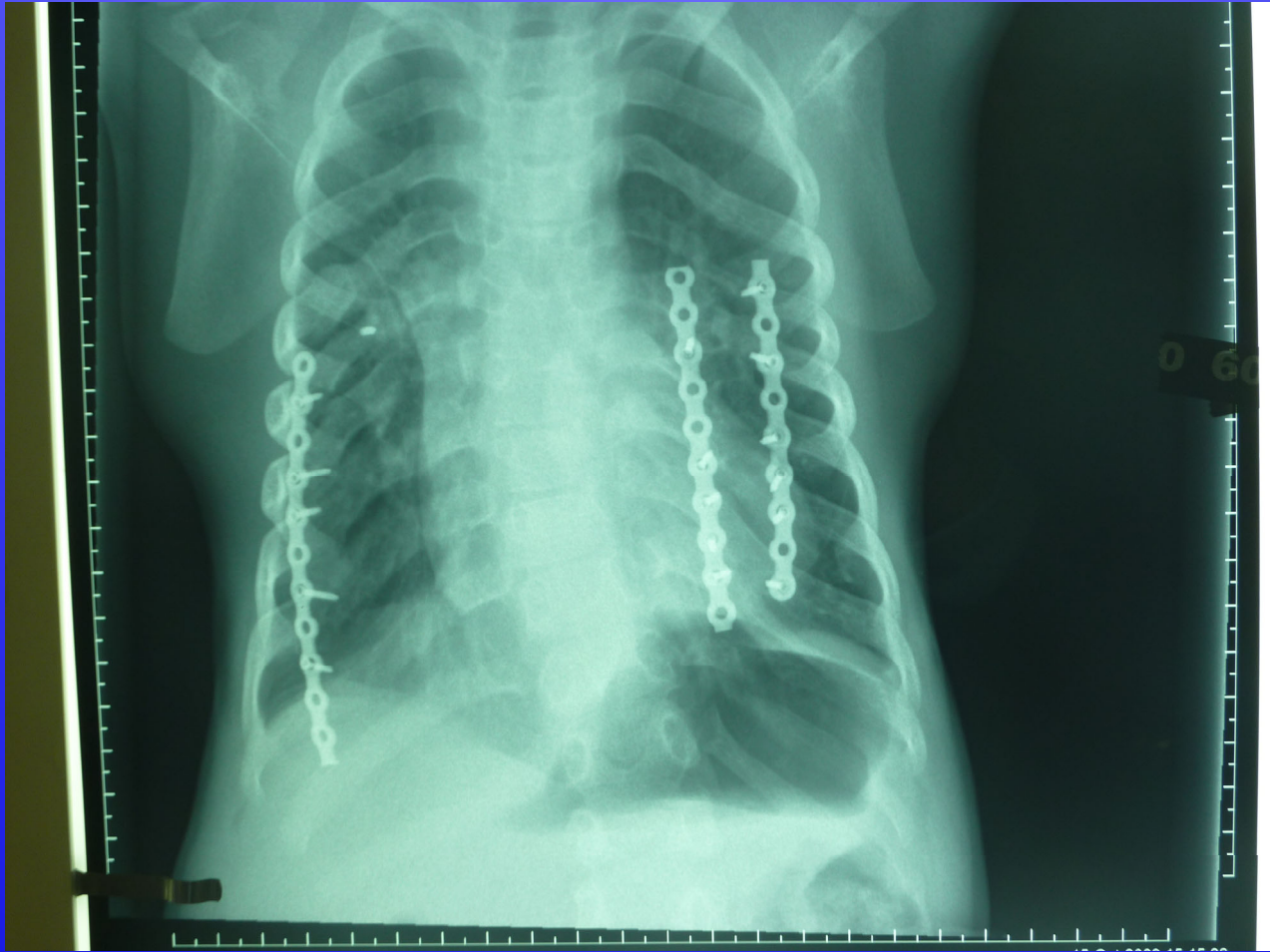


Rib distraction - osteogenesis



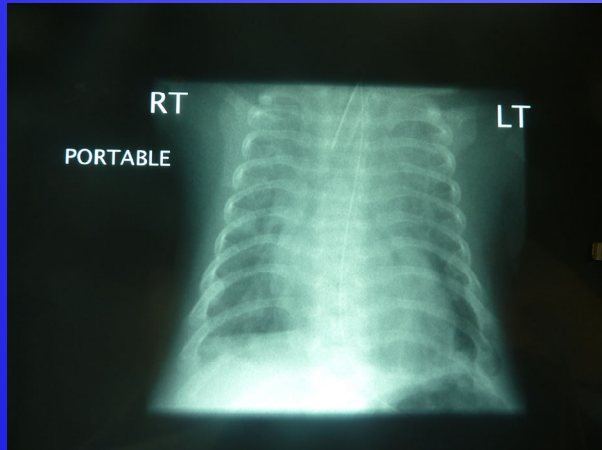
Rib distraction - osteogenesis



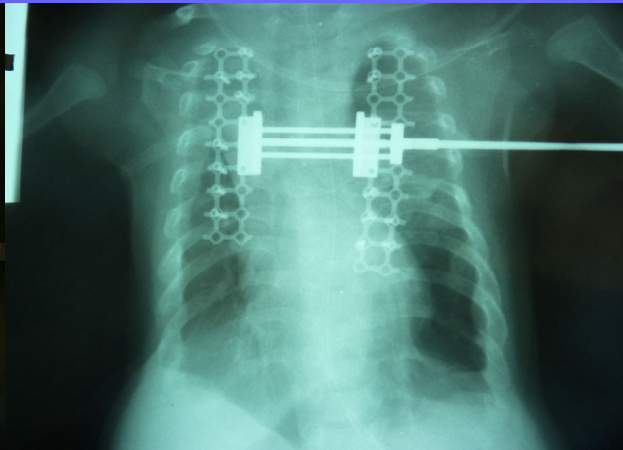


Dynamic thoracoplasty

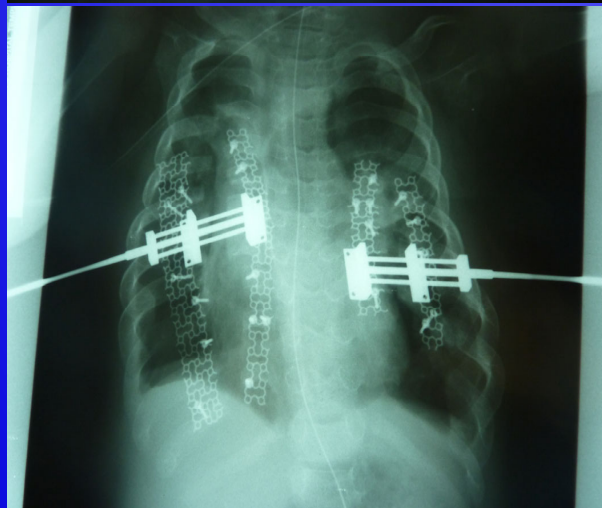
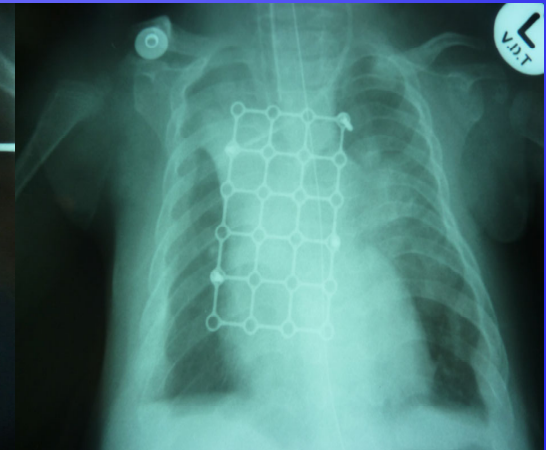
1/12



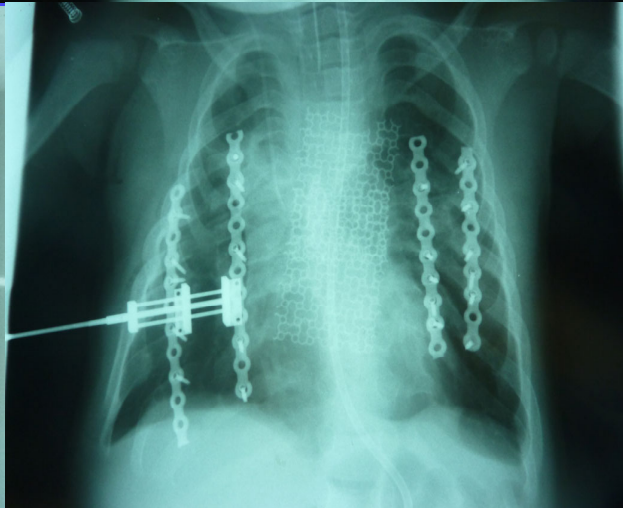
3/12



4/12



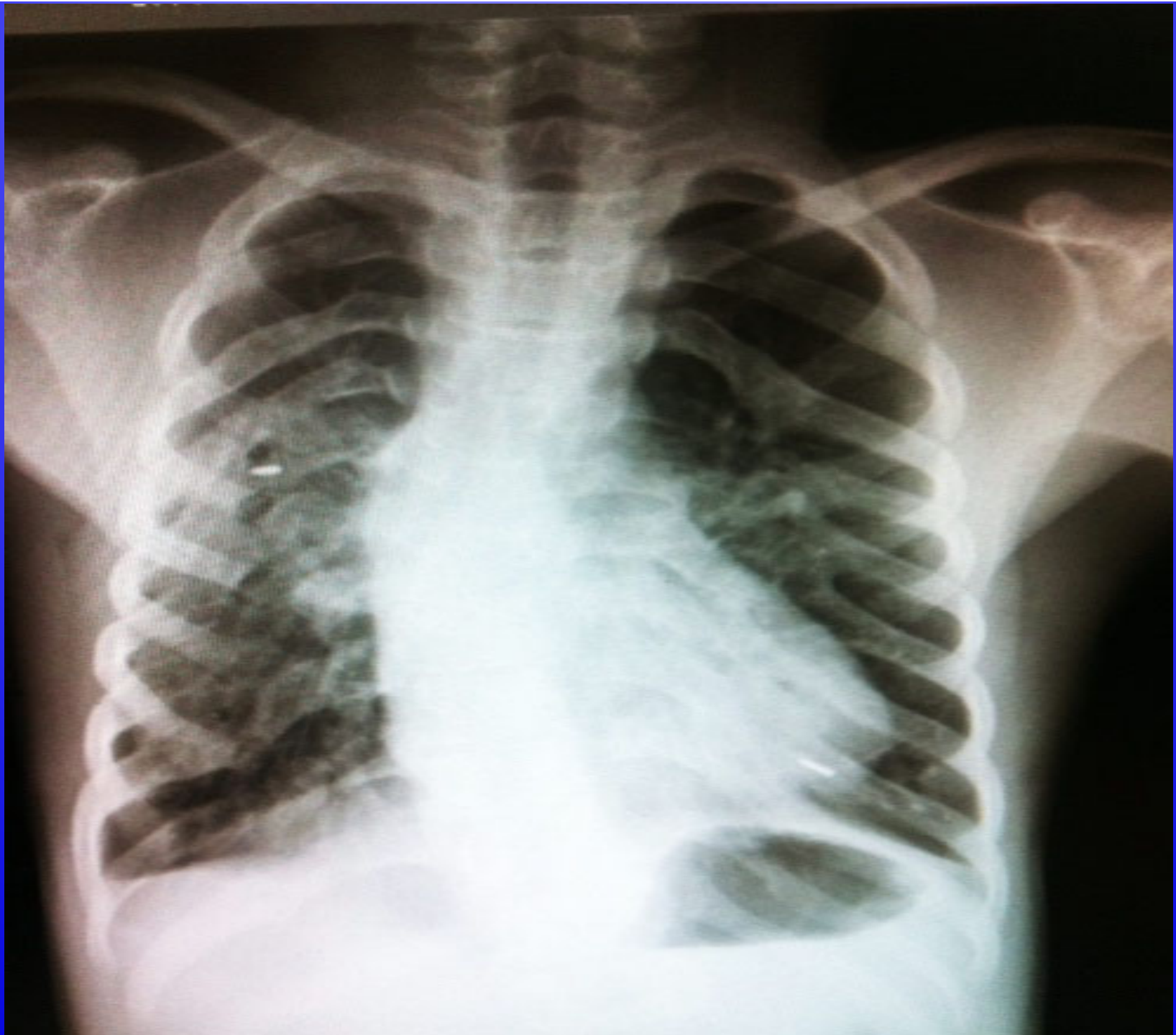
5/12



9/12



3.5 years



“Dynamic Thoracoplasty”

- Life saving in the neonate with severe Jeune syndrome
- Gradual distraction avoids skin /wound problems
- X 10 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

