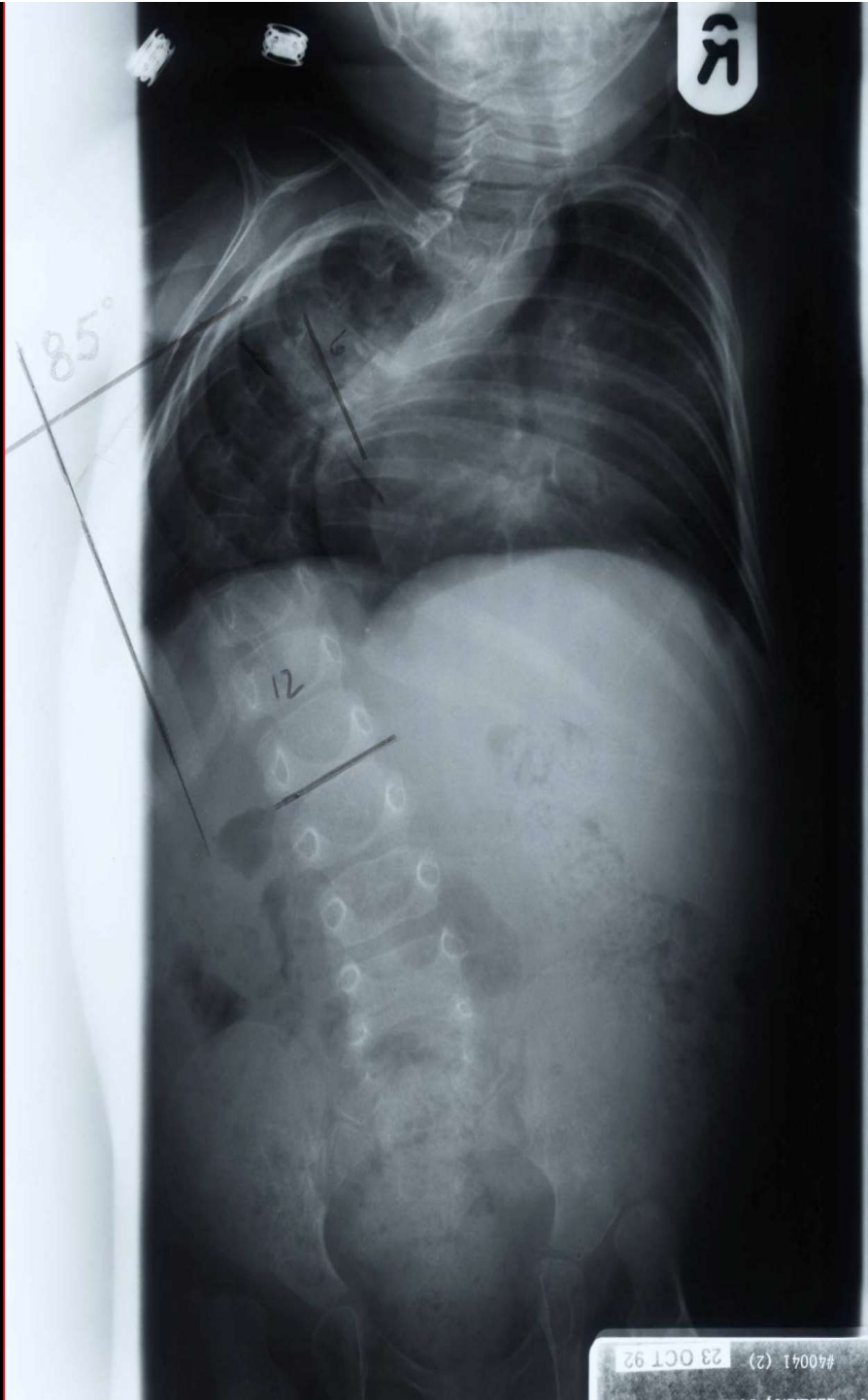


# We Definitely Know the Outcome of What We're Doing (Not)

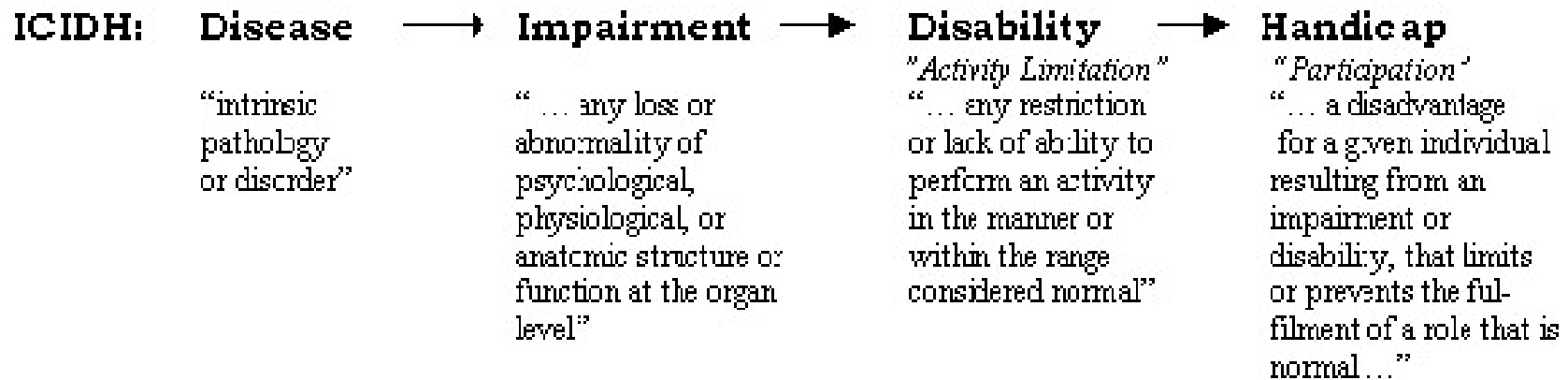
## A call to Action

**SickKids**

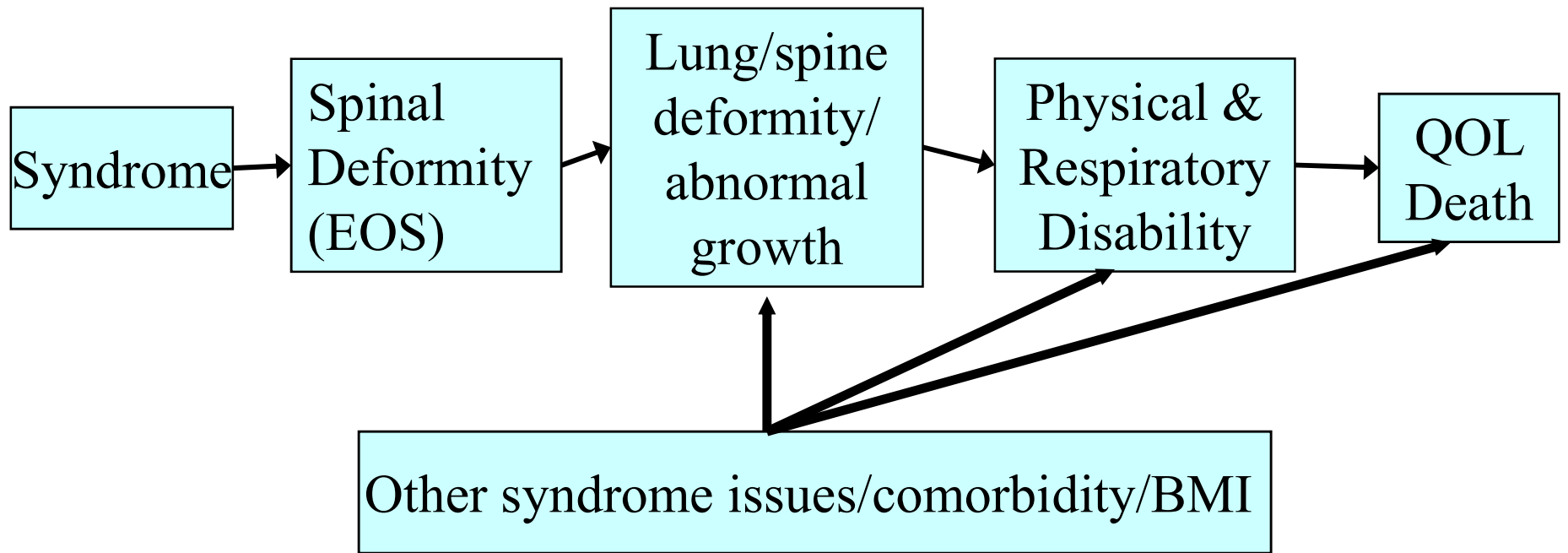




# International Classification of Impairment Disability and Handicap; International Classification of Functioning



Intrinsic Extrinsic  
Factors



# Need to Define Clinical and Anatomic Abnormalities

- Define clinical clusters; eg arthrogyryposis or thoracic insufficiency
- Define diseases; eg Jarcho Levin vs spondylcostal dysplasia and spondylothoracic dysplasia

# Potential outcome measures

- **Anatomic**: ribs, vertebrae, muscles
- **Chest**: size, shape, compliance
- **Radiographic**: Cobb, interpedicular line ratio, decompensation, shoulder level, rotation, posterior hemithorax symmetry ratio, sagittal costophrenic depth ratio, spinal penetration index, thoracic height & depth, abdominal lung positioning, space available for lungs
- **CT**: lung area
- **PFT**: volumes, ABG/saturation, DLCO, assisted ventilation rating
- **QOL**: CHQ-PF50

# Treatment May Worsen Prognosis Short or Long Term

- Early complications; infection, neurologic compromise, skin breakdown, death
- Late complications; fusion, chest wall stiffness, instrumentation or fixation failure

# What has been published

- Early fusion associated with worse PFTs (Goldberg 2003)
- CT lung volumes related to PFTs; shorter, higher, more rotated curves associated with lower CT volumes, worse R>L (Adam 2007)
- More extensive fusions and more proximal fusions associated with worse PFTs (Karol 2008)



# Is it working

- VEPTR for spondylocostal dysplasia in 20 patients stabilized or improved AVR (Ramirez 2010)
- Sternoplasty and rib distraction allowed child with Jeune syndrome to live for >30 months (Conroy 2010)

# What do We Need to Do

- Establish minimal and standardized measures: (formal endorsement, consensus process, periodic revision)
- Develop needed measures: e.g. disease-specific health measure
- Examine inter-relationship of variables
- Establish what's normal

# Next Steps

- Cross sectional studies
- Standardization terminology / measurement
- Sharing of innovations / social media

# Conclusions

- Exciting time with lots of innovation
- Time to bring some order