

Multiple breath inert gas washout in infants and toddlers with cystic fibrosis – methodology and clinical utility.

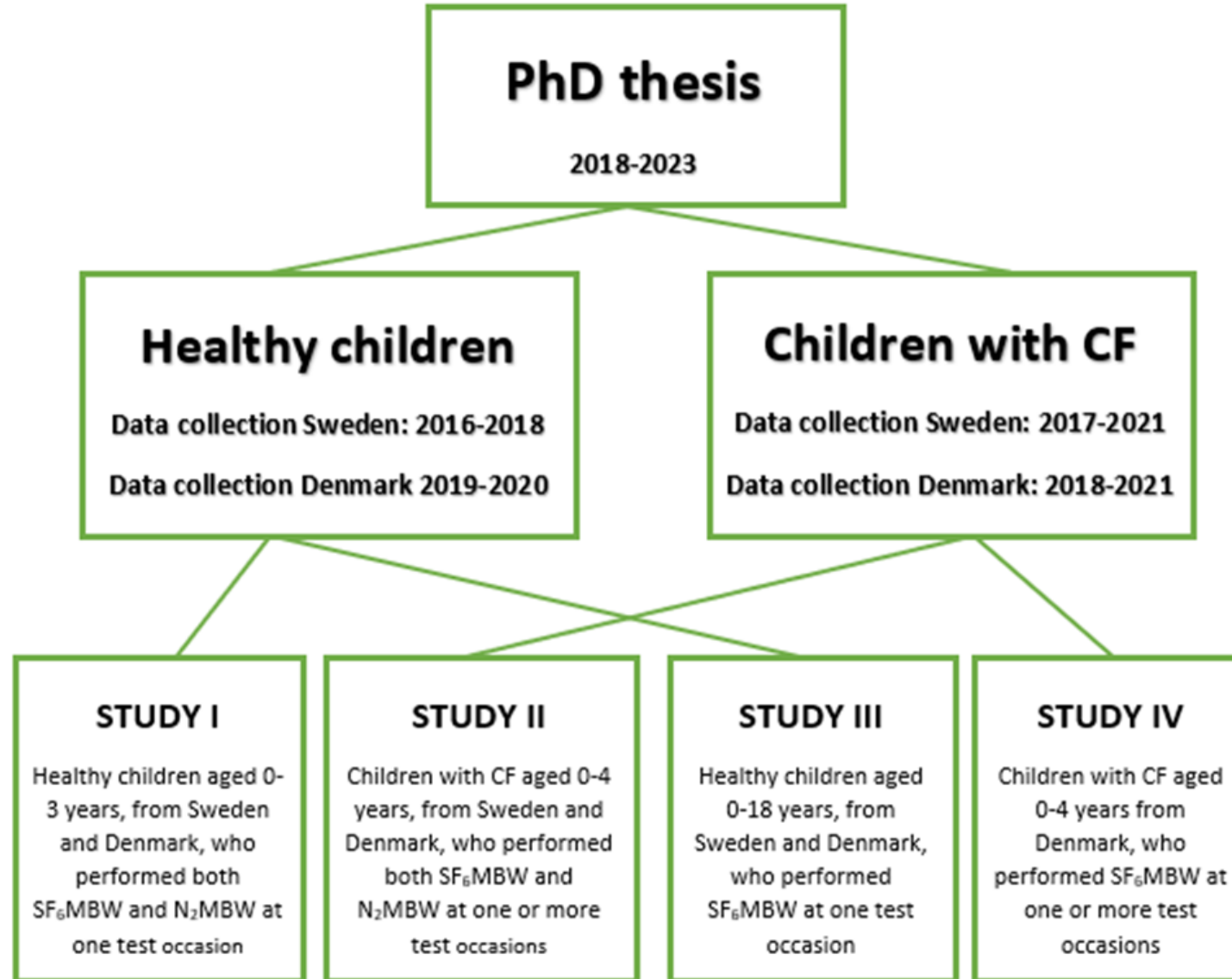
Ventilation distribution efficiency – a new improved outcome



Rikke Mulvad Sandvik, MD

Department of Paediatrics and Adolescent Medicine,
Copenhagen University Hospital, Rigshospitalet, Denmark

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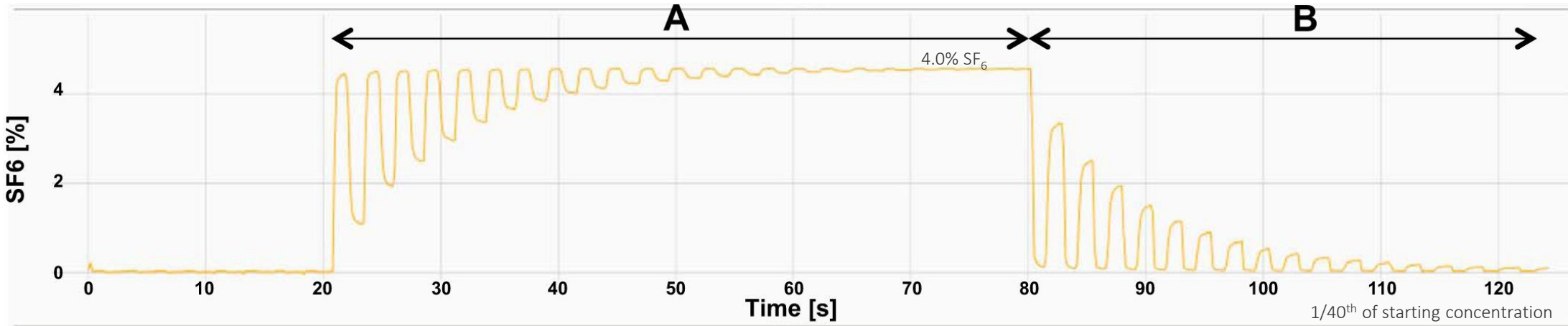


Multiple breath washout

- Tidal breathing
- During sleep
- Can be performed at all ages including infancy
- Primarily assess ventilation distribution inhomogeneity
- It is known to be very sensitive to mild CF lung disease
- And more correlated to structural lung disease assessed by CT or MRI compared to spirometry



SF₆ MBW



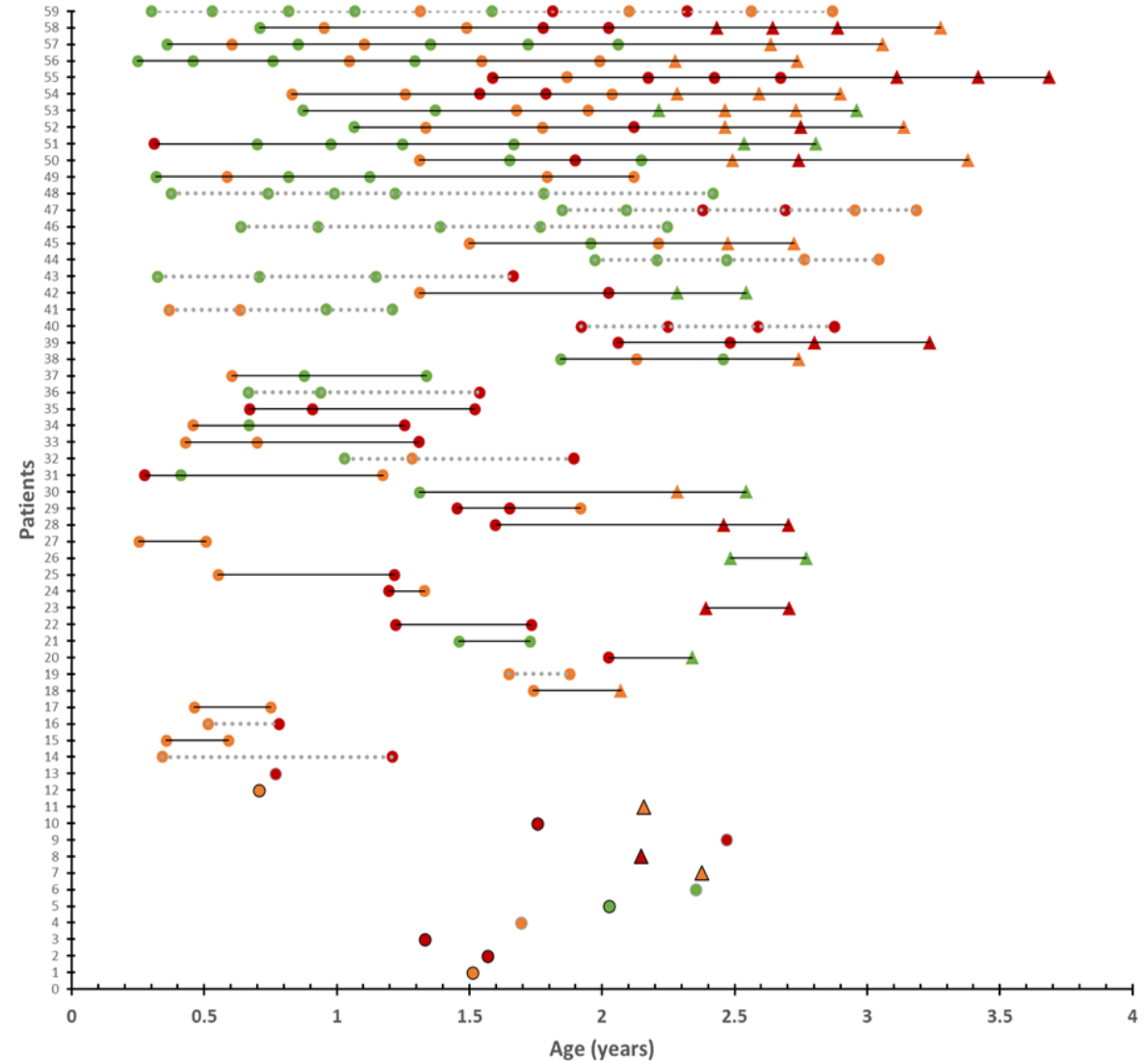
~~LCI = CEV / FRC~~

VDE = FRC / CEV

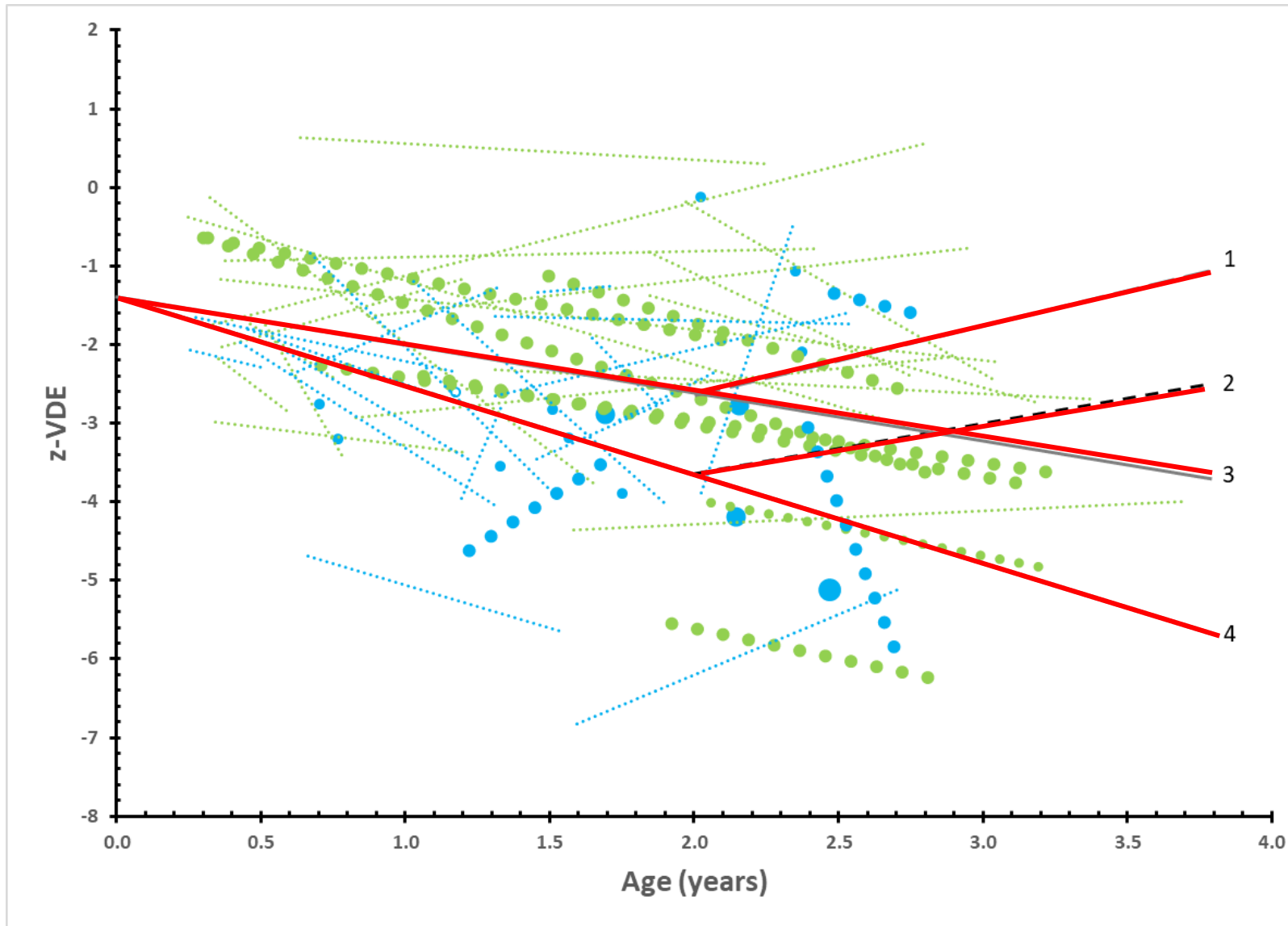
STUDY IV - Use of SF₆ MBW for clinical monitoring

Study cohort:

- 59 children with CF aged 0-4 years
- 211 SF₆ MBW test occasions
- 1512 respiratory secretion samples (average 10/year/child)
- 13 patients were ever infected with *Pseudomonas aeruginosa*
- 22 children started treatment with lum/iva (Orkambi) at age 2 years during the study period



z-VDE progression, effect of PA and Orkambi



Thin trendlines = never PA

bold trendlines = colonized with PA.

Single dots = children with only 1 MBW

green = Copenhagen,

blue = Aarhus

1) mean z-VDE for PA never, plus Orkambi

2) mean z-VDE for PA ever, plus Orkambi

3) mean z-VDE for PA never, without Orkambi

4) mean z-VDE for PA ever, without Orkambi

Ventilation distribution inhomogeneity assessed by MBW demonstrated

- An **overall annual progression**
- The progression rate **significantly worsened** among patients **ever colonized with PA vs. those never colonized**
- An overall **significant beneficial effect** was seen after starting the CFTR modulator treatment (**Orkambi**), leaving this treatment group with **stable lung disease**
- Using **z-scores of VDE for monitoring lung function** in the Danish cohort of young children with CF seemed to provide **clinically meaningful results**.

Clinical monitoring of patients with CF



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