

Pulmonary Care in Duchenne Muscular Dystrophy

Parent JOIN THE FIGHT.
END DUCHENNE.
Project
Muscular
Dystrophy

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Disclosures

- No conflicts of interests



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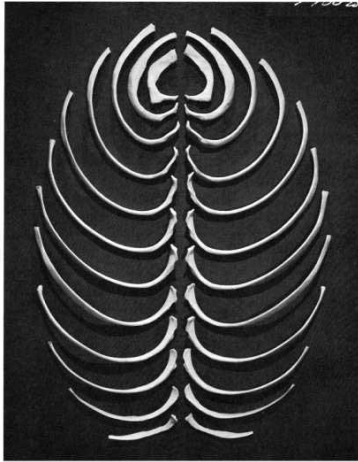
Agenda

- Normal Breathing
- Natural History (Pulmonary)
- Current Recommendations
- Concerning Situations

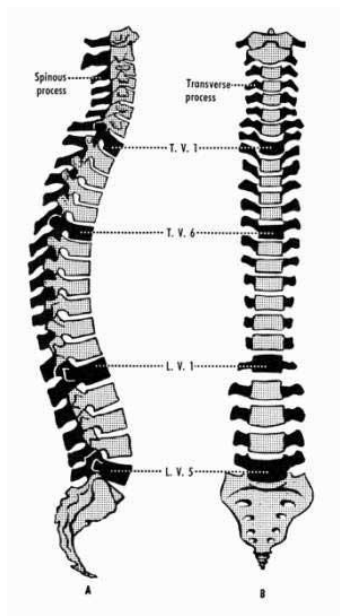


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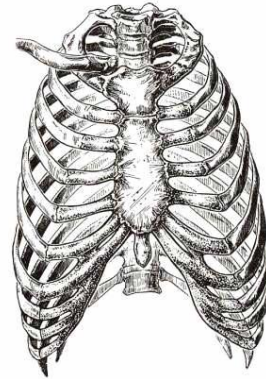
Basic Relationships



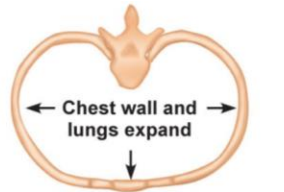
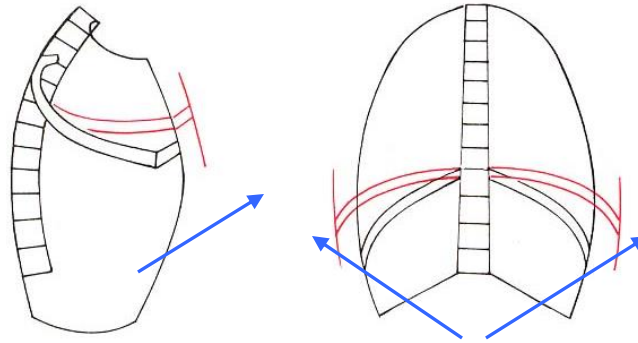
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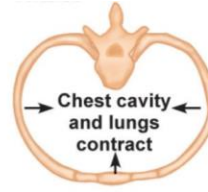
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Normal Breathing



Expansion of ribs moves sternum upward and outward



Ribs and sternum depress

Pulmonary Measures

- Clinical Assessments
- Pulmonary function testing
- Sleep studies



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Clinical Assessments



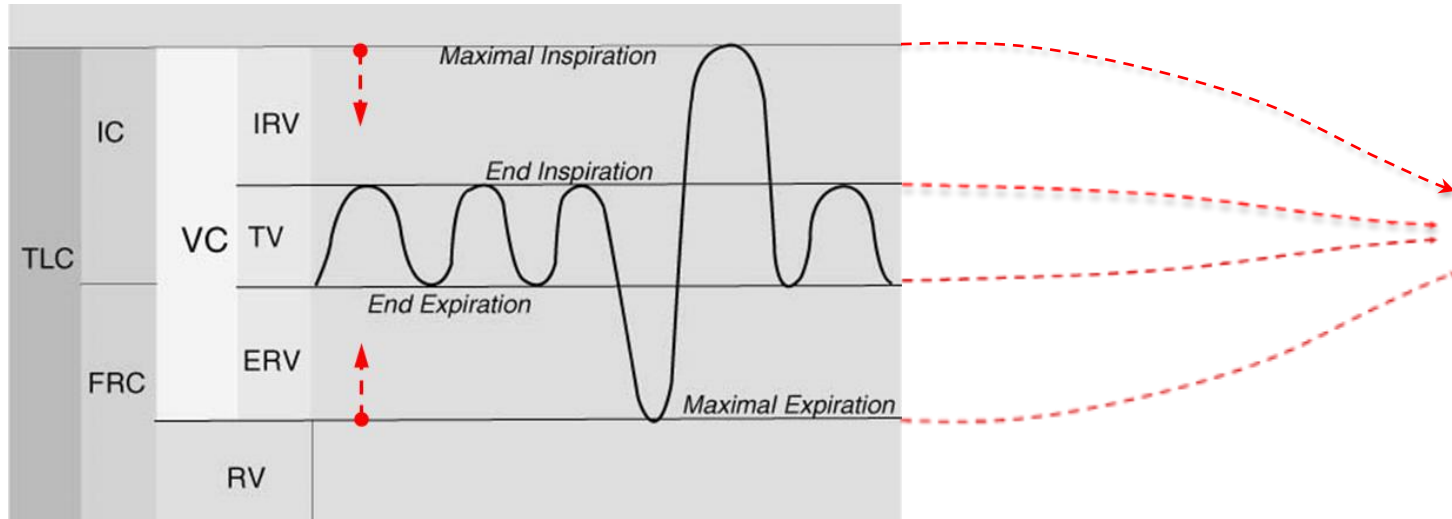
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Clinical Assessments

- Spirometry: FVC
- Respiratory muscle strength
- Peak cough flow
- End tidal CO₂ and respiratory rate

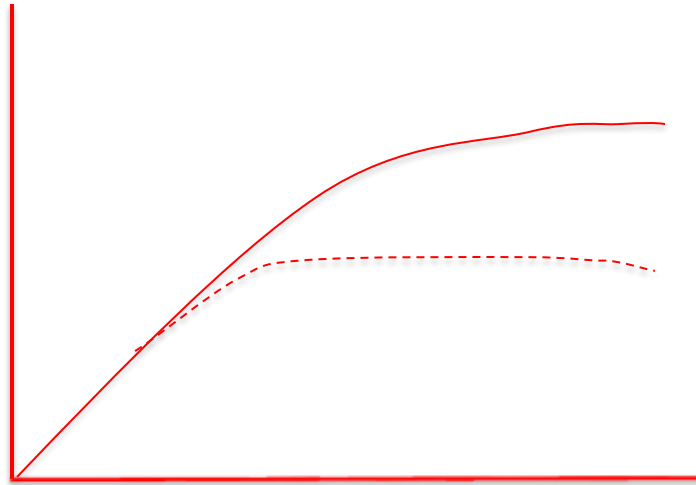


Pulmonary Function Testing



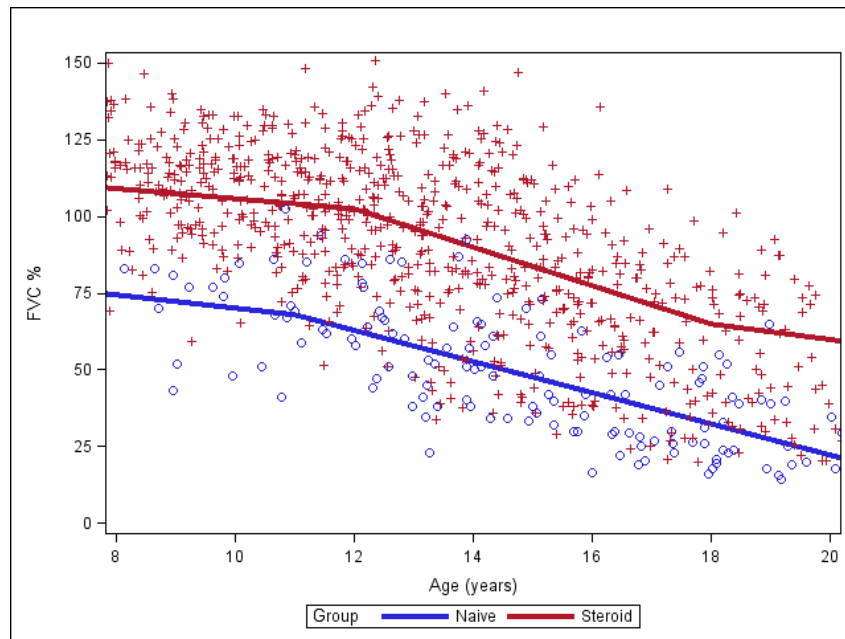
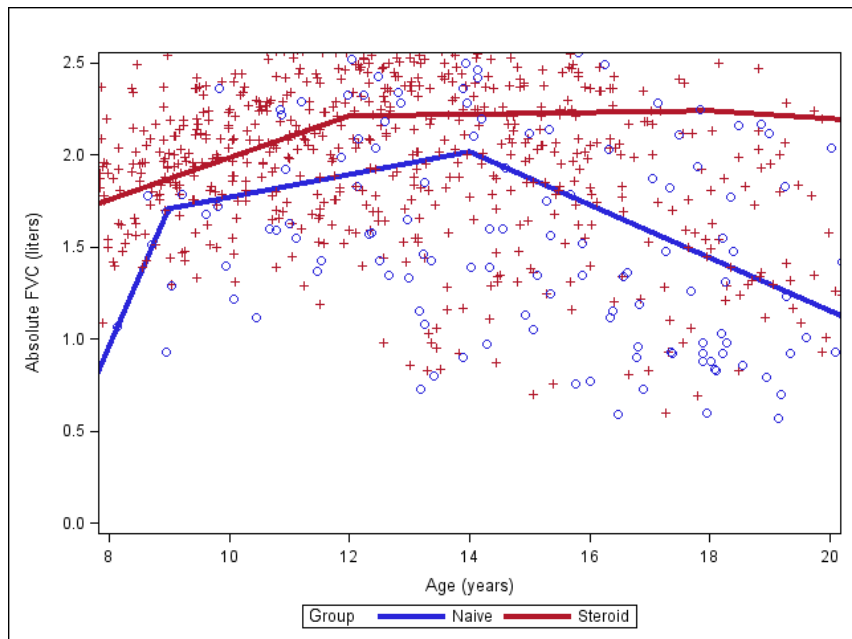
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FVC (L) vs FVC%



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Natural History: GC+ vs GC-



Peak FVC data for Naïve and Glucocorticoid treated DMD patients

	GC-Treated	GC-Naïve	
Measure	Mean±SD	Mean±SD	p-value
Age at Peak Absolute FVC (y)	13.5±3.0	14.3±2.8	0.03
Absolute FVC Peak (L)	2.4±0.6	1.9±0.7	<0.0001
Age at Peak FVC% (y)	11.9±2.9	13.6±3.2	0.002
FVC% Peak	105±25.1	56.0±20.9	<0.0001



PULMONARY MANAGEMENT

	AMBULATORY	EARLY NON-AMBULATORY	LATE NON-AMBULATORY
ASSESSMENTS	<p>Once yearly: FVC</p> <p>Sleep study with capnography* for signs and symptoms of obstructive sleep apnea or sleep-disordered breathing</p>	<p>Twice yearly: FVC, MIP/MEP, PCF, SpO₂, PetCO₂/PtcCO₂</p>	
INTERVENTIONS	<p>Pneumococcal vaccines and yearly inactivated influenza vaccine</p>	<p>Lung volume recruitment when FVC \leq 60% predicted</p>	<p>Assisted coughing when FVC \leq 50% predicted, PCF < 270 LPM, MEP < 60 cm H₂O</p> <p>Nocturnal assisted ventilation with back-up rate of breathing (non-invasive preferred) when there are signs or symptoms of sleep hypoventilation or other sleep-disordered breathing**, abnormal sleep study*, FVC \leq 50% predicted, MIP < 60 cm H₂O, or awake baseline SpO₂ < 95% or pCO₂ > 45 mm Hg</p> <p>Daytime assisted ventilation when despite nocturnal ventilation: daytime SpO₂ < 95%; pCO₂ > 45 mmHg or awake dyspnea***</p>



Airway Clearance Therapies



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Sleep Studies

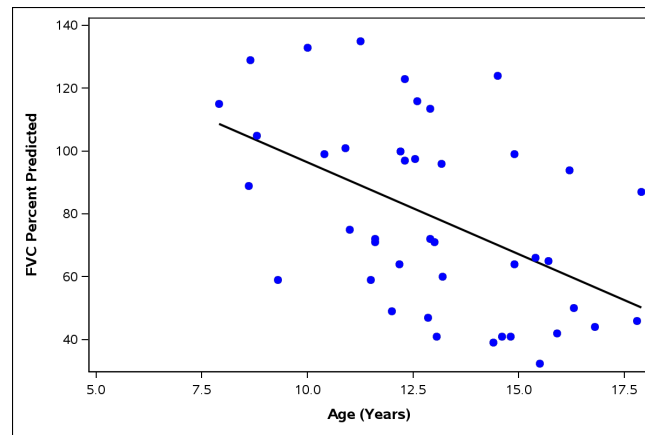
- Indications: Clinical
 - Minority of boys manifest symptoms



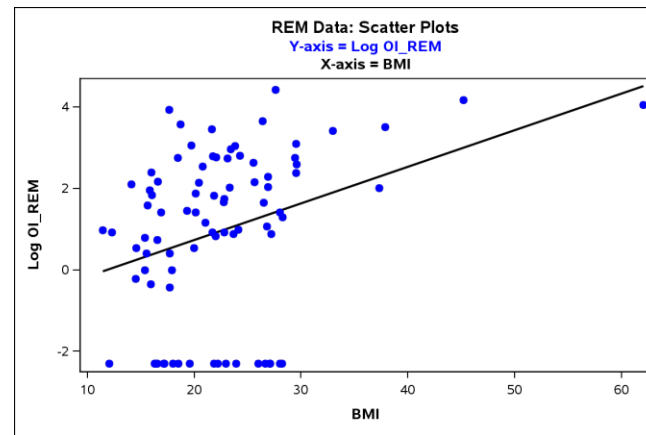
Non-OSA v/s OSA groups

Variable	No (n = 40)	Yes (n = 70)	P-value
Age	10.60 ± 3.08	12.03 ± 2.78	0.014
BMI	20.42 ± 4.98	22.75 ± 7.84	0.093
FEV1%	77.97 ± 36.53	77.62 ± 24.12	0.971
FVC%	79.94 ± 38.75	78.61 ± 25.94	0.897
OI	0.37 ± 0.29	5.24 ± 5.63	<0.0001
OI-REM	2.92 ± 7.03	14.47 ± 17.48	0.0004
PEF%	65.68 ± 27.31	65.39 ± 21.33	0.971
MEP	73.67 ± 35.62	77.85 ± 31.14	0.613
MIP	69.15 ± 29.93	77.61 ± 24.99	0.211
RDI	2.33 ± 3.00	6.46 ± 5.76	0.0001
RDI-REM	7.60 ± 9.63	16.68 ± 17.56	0.004

Regression Model of FVC% and Age



Regression Model of OI in REM sleep and BMI

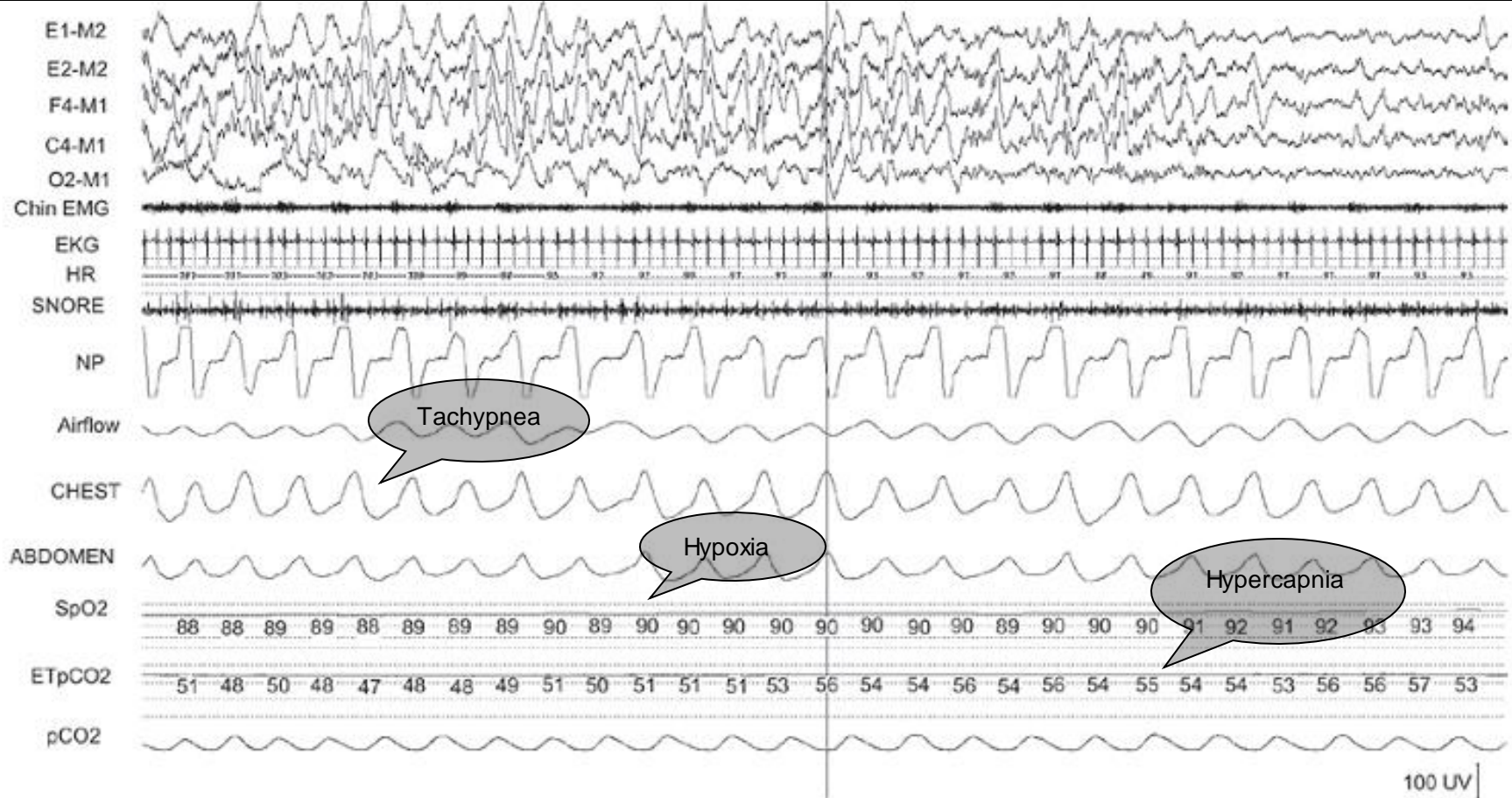


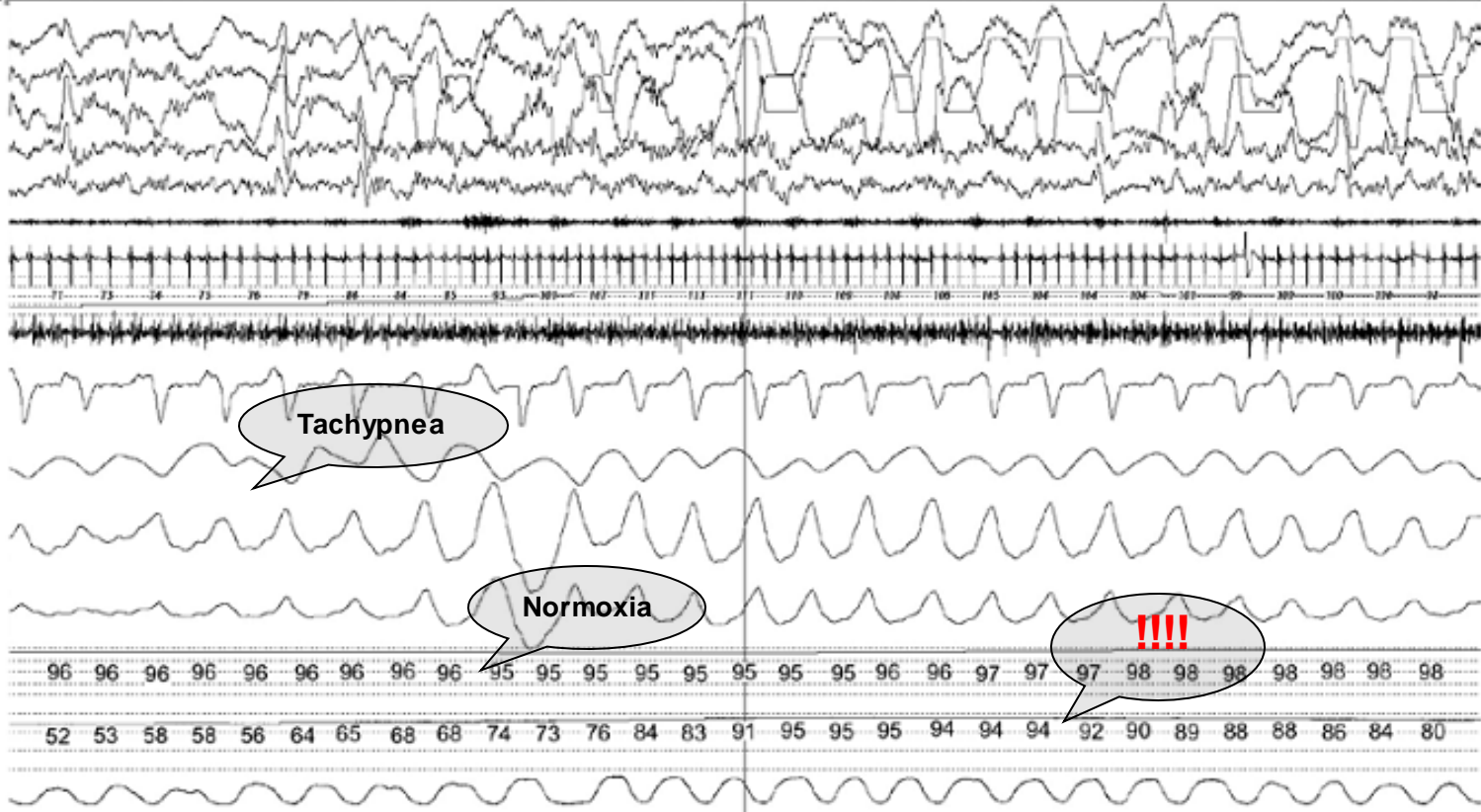
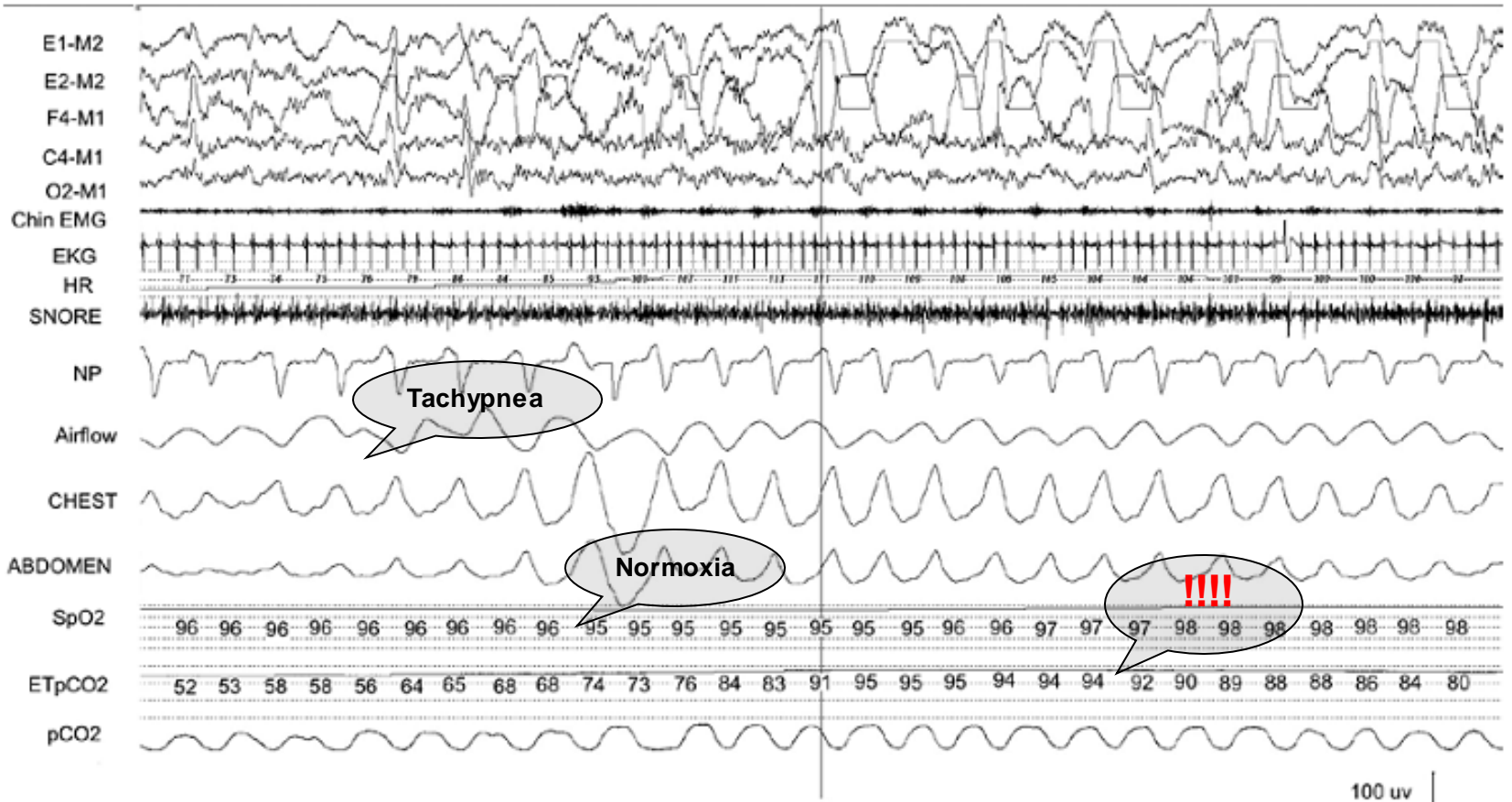
OI during REM sleep positively correlated with BMI ($r=0.33$, $p=0.002$)

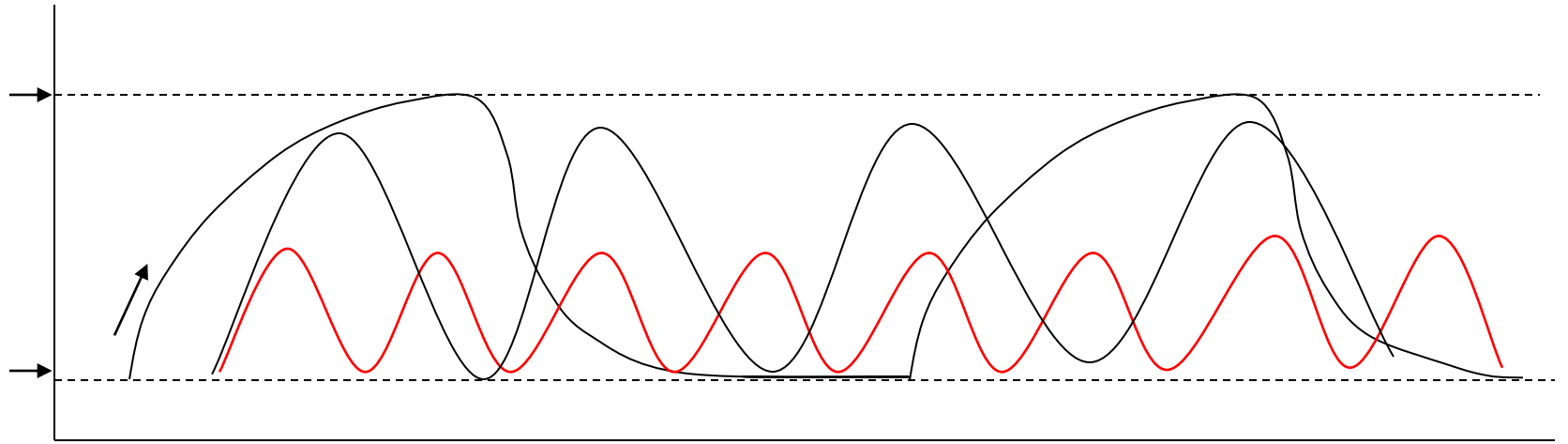
Clinical Situations

- Common colds
- Falls & Fractures
- Why no supplemental oxygen, exclusively?









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Takeaways

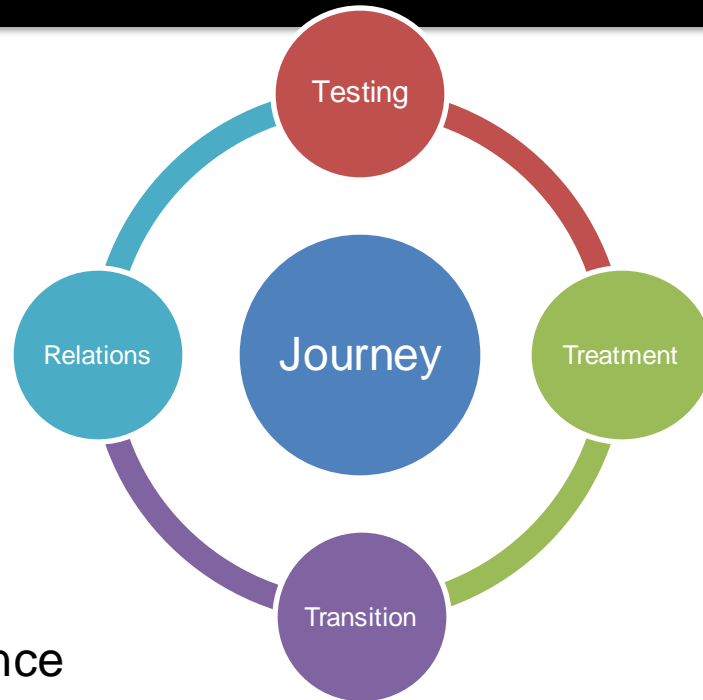
Takeaways

- Early and frequent assessments
- Minimal standards of care; Immunizations
- Special circumstances



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Summarizing



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Thank you!

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Questions?