



**Action Duchenne Conference 2010**  
**Respiratory Care in Duchenne Muscular Dystrophy**  
**Why, When and Where Does it Start?**

**Nicholas Hart**

**Clinical Research Consultant**

**Respiratory & Critical Care Medicine**

**Guy's & St Thomas' NHS Foundation Trust & Kings College London**

**NIHR Comprehensive Biomedical Research Centre**

**Lane Fox Respiratory Unit**

**Department of Critical Care**

**Division of Pain Critical Care & Peri-operative Medicine**

**St Thomas' Hospital**

- **Prior to the introduction of non-invasive home mechanical ventilation (HMV) the mean age of death was 19 years of age**
- **Since the widespread introduction of HMV over 50% of patients survive to 25 years of age<sup>1</sup>.**
- **In addition to living longer, quality of life is maintained by HMV<sup>2</sup>**
- **More recent data suggest that with optimal care, involving cardiorespiratory, nutritional and functional support, a proportion of DMD individuals may live beyond 30 years<sup>3</sup>**

1. Eagle M *et al. Neuromuscul Disord* 2002; 12(10): 926-9

2. Bach JR *et al. Am J Phys Med Rehabil* 1991; 70(3): 129-35

3. Toussaint M *et al. Eur Respir J* 2006; 28(3): 549-55

# **How Does The Lane Fox Respiratory Unit Provide Respiratory Support?**

- **14-Bedded Purpose-Built Unit**
- **2 Purpose-Built Outpatient Rooms**
- **1 Ward Manager**
- **42 Nurses**
- **2 Outreach Nurse Practitioner**
- **3 Respiratory Physiotherapist**
- **1 Rehabilitation Physiotherapist**
- **4 Technicians**
- **1 Neurology Consultant**
- **2 Rehabilitation Consultants**
- **4 Respiratory Consultants**
- **1 Cardiology Consultant§**
- **3 Spinal Surgeons\***
- **1 Orthopaedic Surgeons from Gait Disorder Service\***

§ Cardiothoracic Unit \*Evelina Children's Hospital

# **What Respiratory Support Does Lane Fox Respiratory Unit Provide?**

**DMD Patients**

**Longterm Respiratory Support**

**Acute Respiratory Support**

**Respiratory Assessment**

**Establish & Service  
HMV**

**Regular Reviews**

**Nutritional Assessment  
PEG Insertion**

**Pre-op Assessment  
Post-op Support**

**Assess for Cough  
Assist Device**

**Emergency Admission  
To Lane Fox Unit**

**Non-Invasive Ventilation**

**Respiratory  
Physiotherapy Support**

**Liaise with  
AICU & PICU**

**Liaise with  
Local Hospital**

**Weaning from Ventilation/  
Removal of Tracheostomy**

# DMD Patients

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## Acute Respiratory Support

Emergency Admission  
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## Home Care Plan for Chest Crisis Management

### Symptoms to watch out for

- Chesty cough
- Fever
- Sleep disturbance night or sleepy in day
- Lack of appetite
- Feeling 'under the weather'

### What to do straightaway

- Check temperature
- If > 37.5 treat with paracetamol and contact GP to start antibiotic treatment
- Use cough assist +/- chest physiotherapy
- Use mask ventilator as well if breathless
- Telephone Lane Fox Respiratory Unit

### Move to second line management if any of the symptoms listed below:

- Increasing use of mask ventilator
- Unable to breathe properly when off it
- Fast, shallow or deep gasping breathing
- Poor fluid intake
- Increased secretions or inability to clear these with cough assist +/- chest physiotherapy
- Yellow/green sputum
- Blue hands or lips
- Rapid decline in breathing
- Difficulty talking in full sentences
- More sleepy than normal

### 2<sup>nd</sup> line management

- Contact Lane Fox Respiratory Unit on 020 7188 3435 or 020 7188 3434 OR
- Contact Emily Ballard, Transitional Care Coordinator on 0207 188 8069 or bleep 2433

**If urgent call the switchboard on 0207 188 7188 and ask for the 'Nurse Advisor Lane Fox Unit' on bleep 1572 who is available 24 hours a day**

### 3<sup>rd</sup> line emergency management (unconscious or severe distress)

- Call 999 for admission to local hospital
- Use mask ventilator in ambulance
- Ask A&E staff to call LFU on arrival

**DMD Patients**

**Longterm Respiratory Support**

**ICU Admission  
Tracheostomy  
EOLC**

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**What Innovative Strategies Have Been  
Employed in the Lane Fox Respiratory Unit?**

# **Neuromuscular Transition Service**

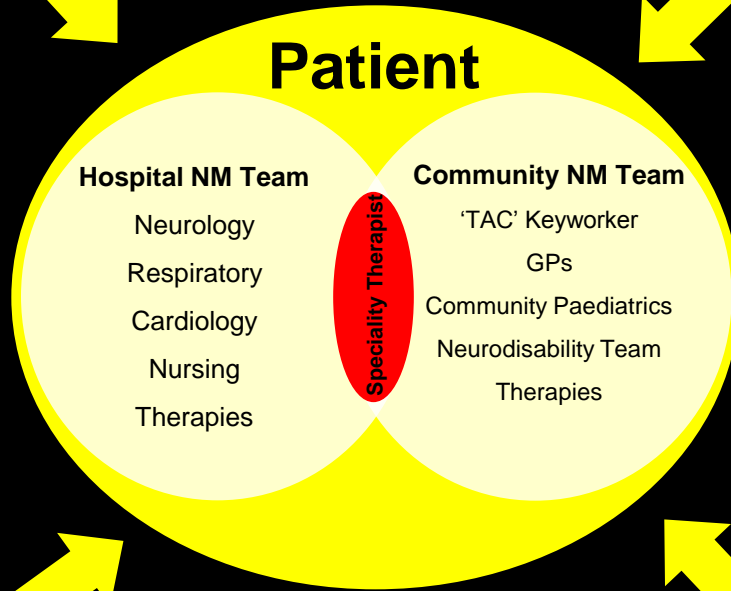
**Guy's & St Thomas' Charity**  
**New Service Innovation Grant**  
*(Project Code: G080109)*

**Database development**  
Demographics  
Cardiac  
Respiratory  
Functional

**Patient & parent/carer Education**  
Introduction to devices

**Community support**  
Identify patients "At risk"  
Admission/prevention  
Facilitate early discharge  
Provide family/ carer support  
Liaison with school

**Yearly cardio-respiratory assessment**  
FVC, PCF  
ABG  
MIP, MEP, SNIP  
Nocturnal Oximetry & Capnography  
ECG & Echocardiogram



**End of life care**  
Liaison with palliative care

**Nutrition assessment**  
Weight  
BMI  
PEG Insertion

**Streamlining between paediatric & adults services**  
Liaison between hospital & community  
Liaison between hospital specialities  
Liaise and support PICU  
Liaise with other units

**Function & mobility maintenance**  
Maintain Strength  
Prevent Contractures  
Promote Independence

**Outpatient reviews**  
Initiation of NIV & Cough Assist Machine

# **Are Respiratory Crises Preventable?**

- **We hypothesised that respiratory review and intervention during adolescence would reduce the number of respiratory crises and length of hospital stay (LOS)**
- **We analysed the data from medical records on all children and young adult patients under the care of the LFRU within a twelve month period**
- **We identified 39 patients under follow up with an age range of 14 to 36 years**

|  | <b>No previous respiratory review prior to 1<sup>st</sup> respiratory crisis admission</b> | <b>Previous respiratory review prior to 1<sup>st</sup> presentation respiratory crisis admission</b> |
|--|--|--|
| <b>Mean Age (years)</b>  | <b>16 (11-22)</b>  | <b>16 (16-19)</b>  |
| <b>Mean Length of Stay (days)</b>  | <b>19</b>  | <b>8</b>   |
| <b>% Patients requiring invasive ventilatory support and tracheostomy</b>                            | <b>8</b>   | <b>0</b>   |
| <b>Mean LOS (days) for weaning and tracheostomy removal</b>  | <b>36</b>  | <b>0</b>   |
| <b>Mean interval time between last clinic review and presentation as a crisis admission (months)</b> | <b>14</b>  | <b>2.4</b>   |

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- **These data support the need for specialist respiratory review before the 1st respiratory crisis**
- **A respiratory review prior to admission with a respiratory crisis reduces the incidence of intubation, ventilation, tracheostomy formation and prolonged weaning on the ICU as well as the mean LOS**
- **Collaboration between paediatric neurology and specialist respiratory support will ensure timely intervention to reduce the severity of a crisis**
- **We have adopted a transitional care model from paediatric to adult services within our institution and we hypothesise that this will improve outcome further**

# Clinical Consultation

# History

- **Unexplained breathlessness in patients with DMD**
- **Many patients with DMD are often only capable of limited exercise and thus breathlessness can be a late symptom**
- **Severe generalised inspiratory muscle weakness can cause symptoms of nocturnal hypoventilation**
  - **disrupted sleep pattern**
  - **early morning headache**
  - **daytime somnolence and fatigue**
  - **impaired intellectual function**

**What Respiratory Tests Do  
We Carry Out as Routine?**

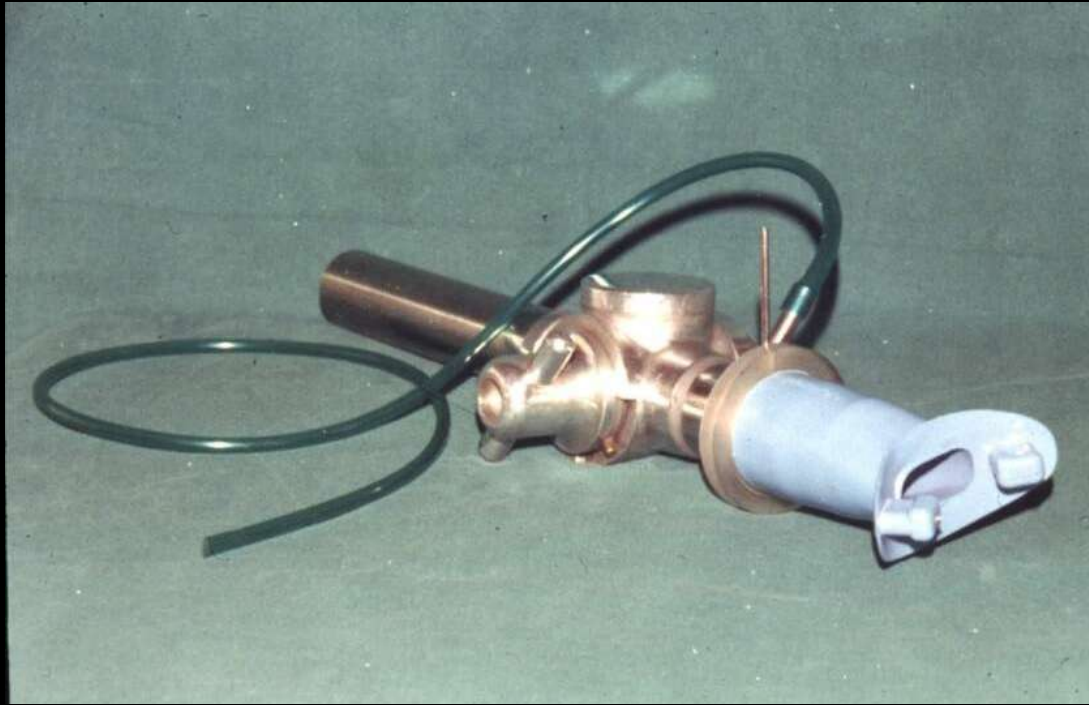
**FVC**

**PI<sub>max</sub>**

**PE<sub>max</sub>**

**SNIP**

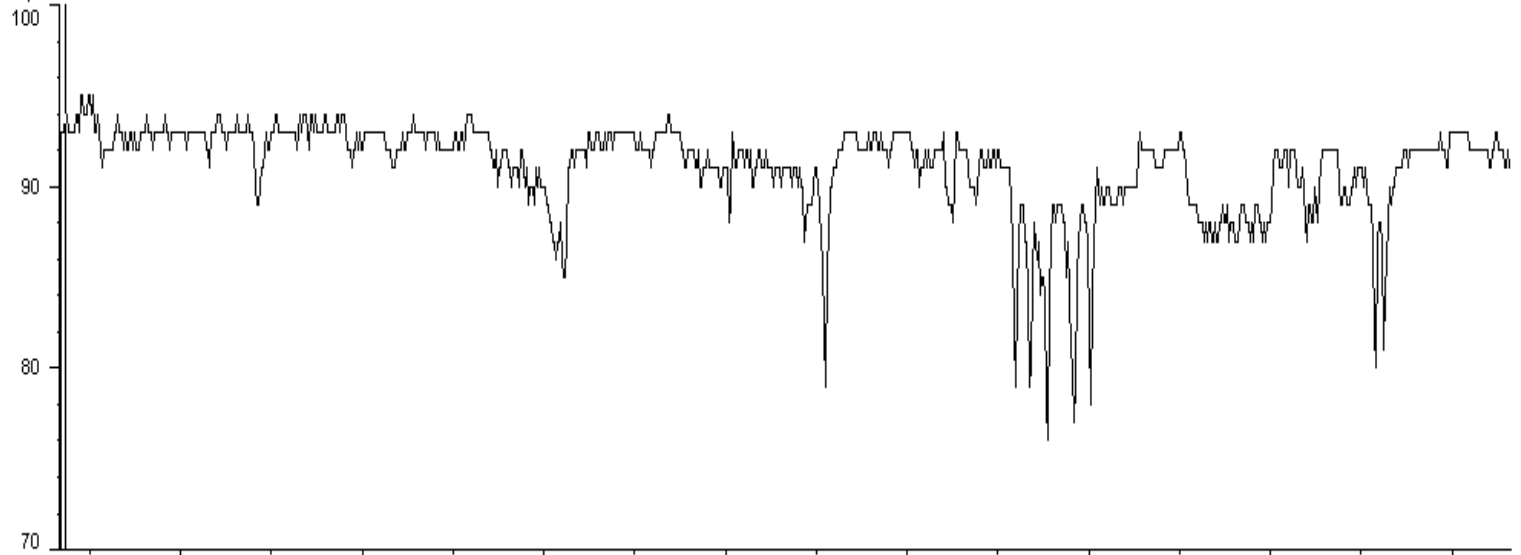
**SpO<sub>2</sub>**



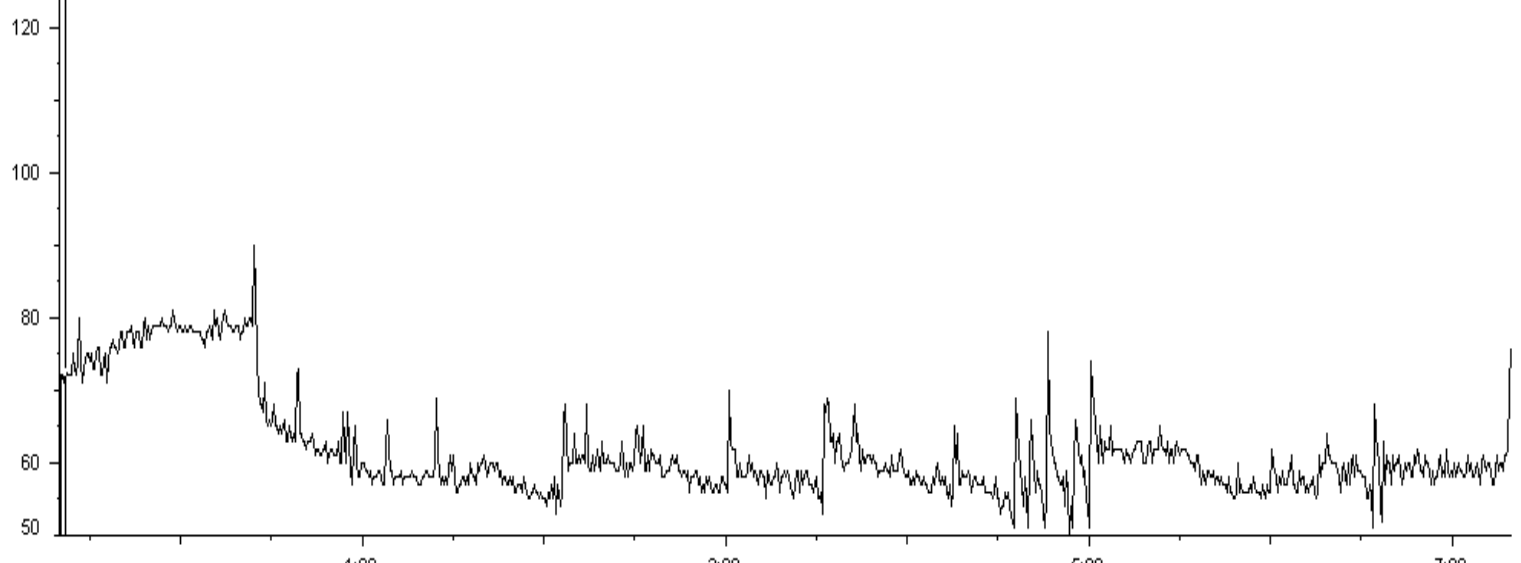


# **Overnight Assessment Oximetry at Home**

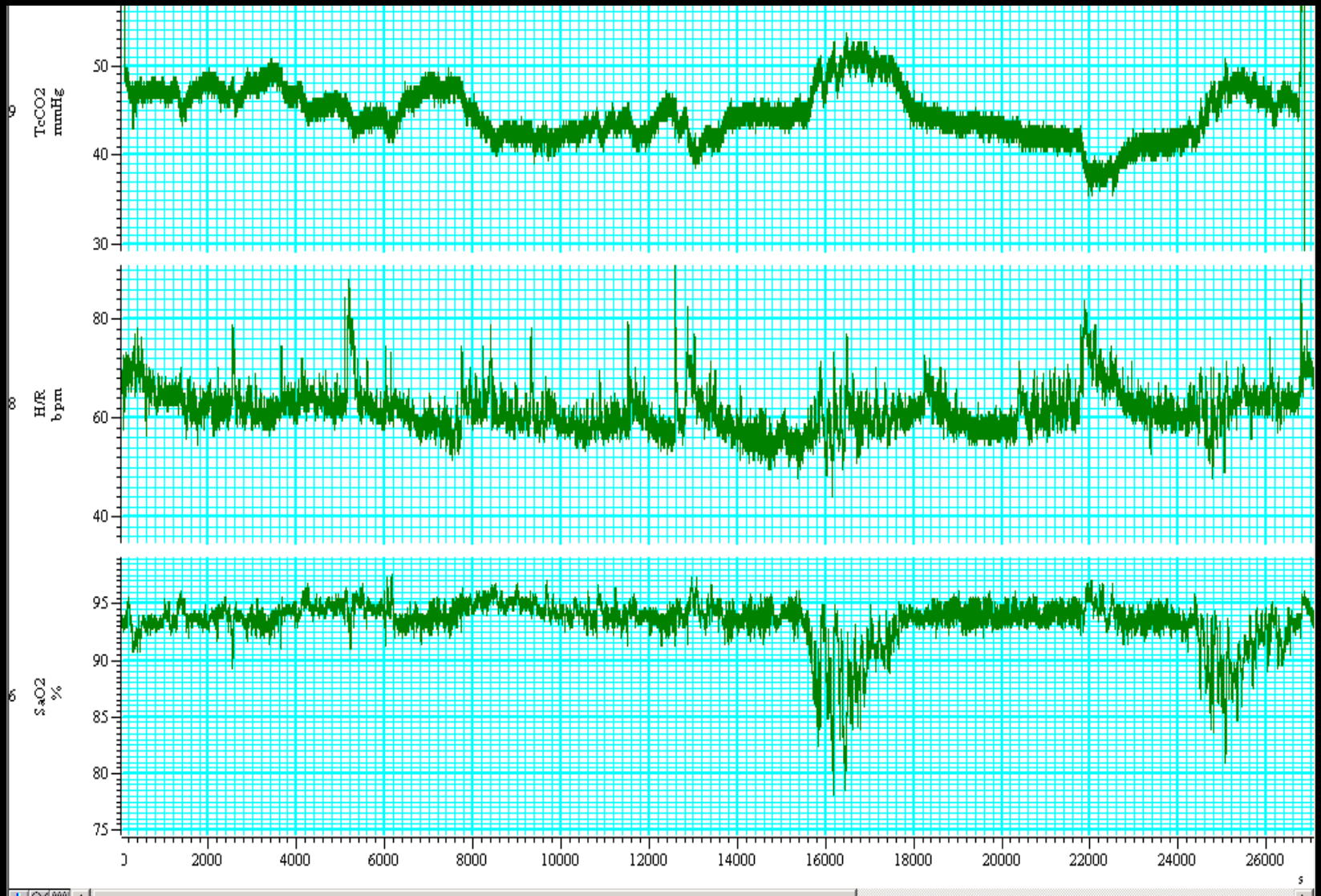
1:00 3:00 5:00 7:00 Saturation 94.00



bpm Pulse 72.00



**Overnight Assessment  
Oximetry & Transcutaneous  
Capnography in Hospital**



# **Management of Nocturnal Hypoventilation**

WARD USE ONLY

For CLINICAL TRIALS ONLY use to be used for Patients  
ALWAYS USE AN OXYGEN/RESERVE ALARM on Tracheostomized Patients





CPAP/PEEP Therapy  
Model No. 4700000000  
© B & D Electromedical

IPAP 28.0cm  
EPAP 10.0cm  
Ti 1.00sec  
Back-Up 188BPM  
Mode PRESSURE CONTROL


Rate 20 BPM  
Ext.Vol. 0.34 L

Hi Alarm 100L/m  
Lo Alarm 20L/m

Pressure 23.7  
Flow 37

Ext. Batt.   
Power   
 

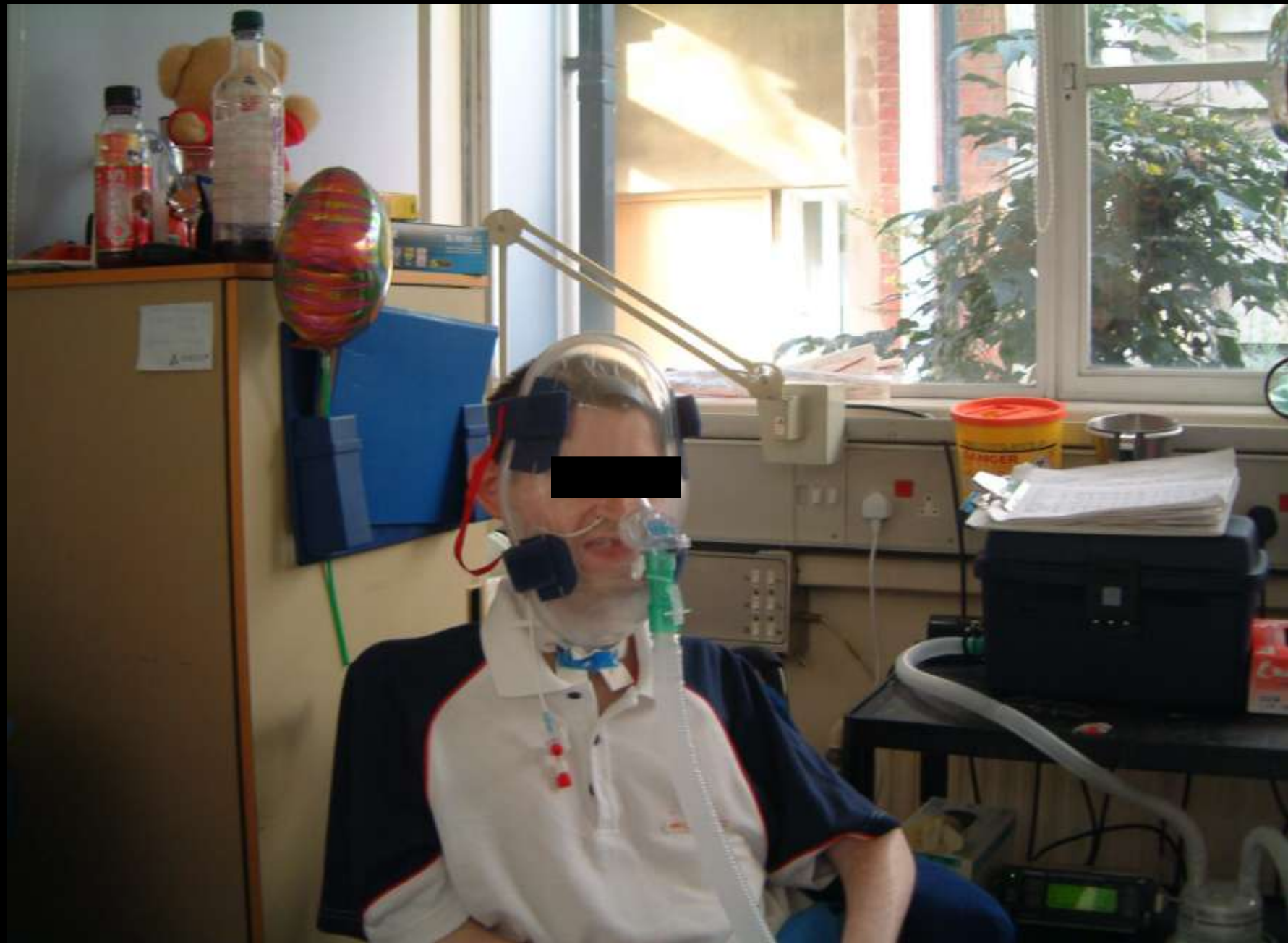
**NIPPV 3**  
B & D Electromedical  
Staford-on-Avon  
England



Handwritten notes on a clipboard, including a patient chart and a list of items:

- Handwritten notes on a patient chart.
- Handwritten list of items:

  - 1. 100% O2
  - 2. 100% O2
  - 3. 100% O2
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# Physiotherapy Techniques

# Insufflator-Exsufflator







# Lane Fox Vision of Healthcare



# Lane Fox Vision of Healthcare



**Fair**  
**Personalised**  
**Effective**  
**Safe**



**The Lane Fox Respiratory Unit  
Management Team**

- **Natalie Grey (Ward Manager)**
- **Kelly Stewart (Outreach Nurse Practitioner)**
- **Alex Curtis (Rehabilitation Physiotherapist)**
- **Emily Ballard (Specialist Transitional Care Coordinator)**
- **Hira Miah (Technical Services)**
- **Robin Howard (Neurology Consultant)**
- **Robin Luff & Julian Harris (Rehabilitation Consultants)**
- **Craig Davidson, Adrian Williams, Hina Pattani & Nick Hart (Respiratory Consultants)**
- **Gerry Carr-White (Cardiology Consultant)**
- **Jonathan Lucas, Tom Ember & Kai Lam (Spinal Surgeons)**
- **Martin Gough (Orthopaedic Surgeon Gait Disorder Service)**

**Any Questions?**