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INDÉPENDANCE ET EXPERTISE POUR LE PROGRÈS DE LA SANTÉ À DOMICILE

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NONINVASIVE VENTILATION CONFERENCE

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Compliance: what should be our target?

Dr Francois ABEL, MD FRCPCH
Paediatric Sleep and Non-Invasive Ventilation
Great Ormond Street Hospital for Children
London, UK

The child first and always

UCL INSTITUTE OF CHILD HEALTH

Great Ormond Street Hospital for Children NHS Foundation Trust

Compliance vs adherence

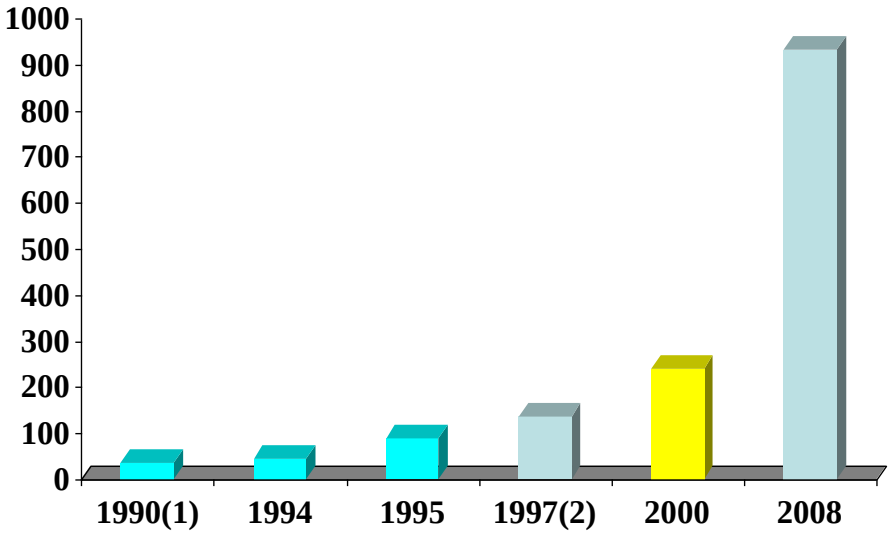
- Compliance connotes unilateral decision-making and expectations that, if the doctor prescribes it, the patient should take it

- Adherence connotes a mutually agreed upon plan between patients/parents and doctor that patients can follow and doctor will support

"It was *worse* than guilty! — The jury found me *noncompliant!*"

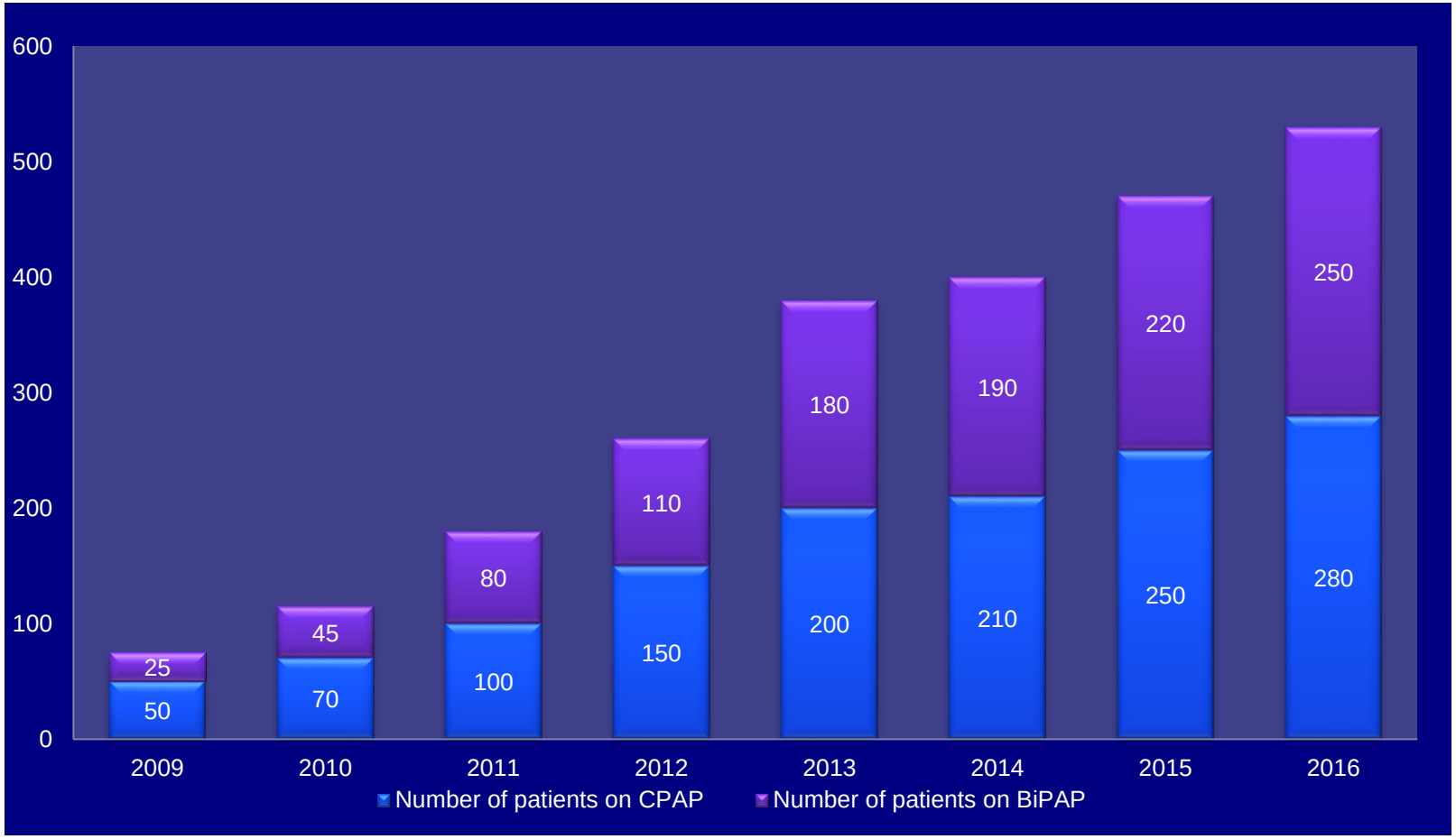
Introduction

- A significant increase in the number of children receiving long term ventilatory support in the UK, most notable in the NIV group, over the past 2 decades[1-3]
- NIV has made significant impact on many conditions: neuromuscular, craniofacial, obesity syndromes, metabolic conditions.
- The exponential increase in both patients and indications for NIV making adherence a key issue



1. *Wallis et al, Arch Dis Child. 2011;96:998-1002*
2. *Robinson, Arch Dis Child. 1990;65:1235-6*
3. *Jardine and Wallis, BMJ. 1999;18:295-9*

Number of patients on LTV 2009-2016



For 2014-2016:

NIV 75%
LTV 25%

Source: Respiratory Sleep Laboratory GOSH

Overview

- What is in the literature about adherence in NIV
  - Factors influencing adherence in NIV
- Lessons from our own cohort
- Wider experience from other centres
- When do we call it a failed NIV case?
  - What are the reasons for failures?
- Lessons learnt: how do we maximise our chances of success? (with the help of NIV CNS and play therapists)

Adherence to NIV

- Adherence to NIV impacts on the efficacy and outcome of treatment for sleep disordered breathing[1]
- Good NIV adherence corrects nocturnal and daytime gas exchange in children with neuromuscular disease[1-2]

Consequences of non-adherence:

  - Acute/recurrent/life-threatening respiratory deterioration

- In adult studies, more consistent use of NIV is associated with better outcomes [3-6]:
  - Reduced sleepiness
  - Increased self reported energy
  - Improved cognitive function

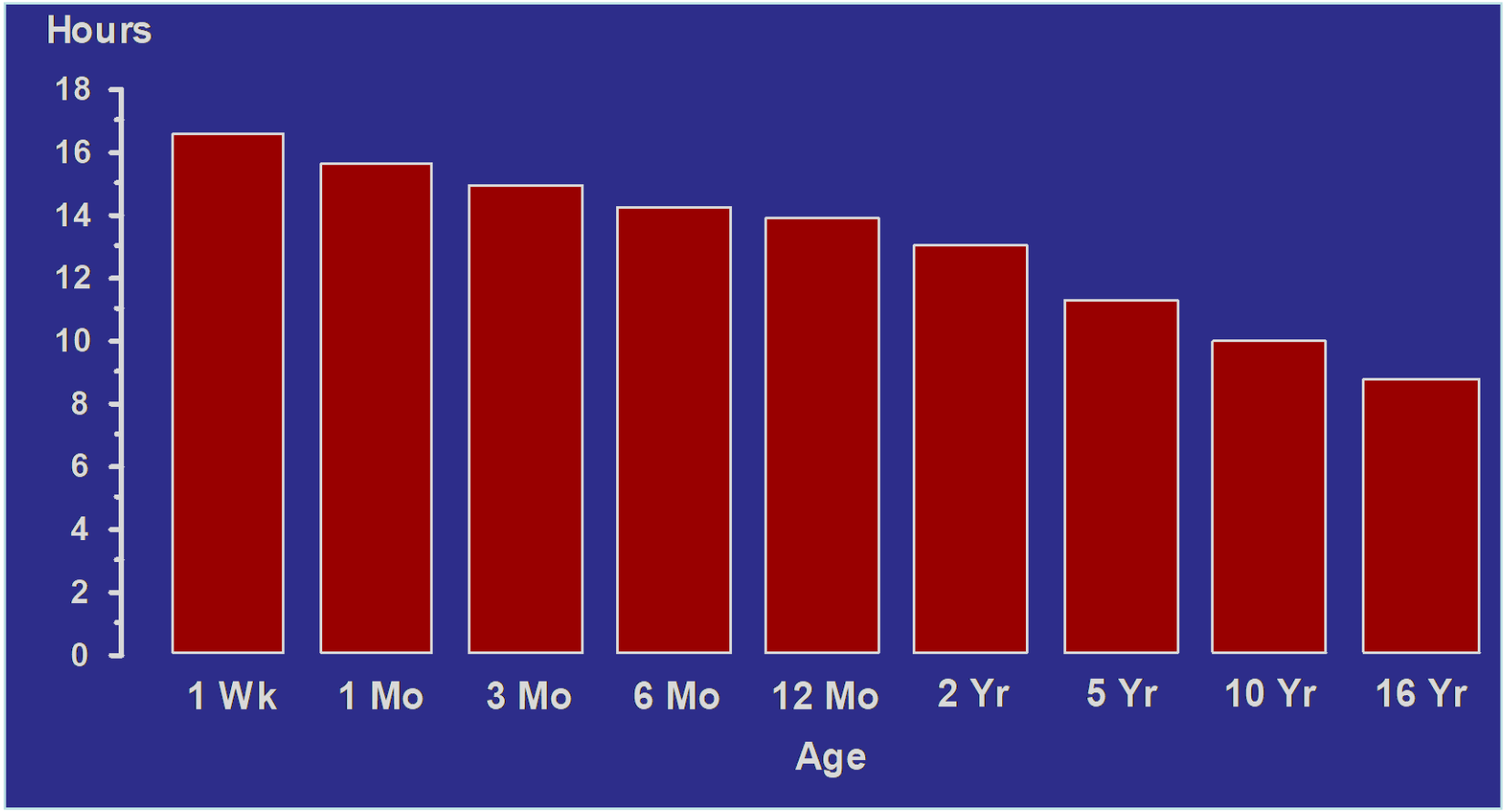
1. *Mellies U et al. Eur Resp J 2003 ; 22:631-6*
2. *Simmons A et al. Eur Resp J 2000 ;16:476-81*
3. *Campos-Rodriguez F et al. Chest 2005 ; 128: 624-33*
4. *Zimmerman ME et al. Chest 2006 ; 130:1772-8*
5. *Weaver TE et al. Sleep 2007 ; 30:711-9*
6. *Dinges DF et al. Behav Sleep Med 2007 ; 5:79-82*

Lessons from adult NIV adherence studies
The “4 hours” rule

- Based on several studies, compliance of ≥4 h per night has been considered acceptable.
- However, dose–response studies have found that different compliance levels achieve different dimensions of clinical improvement [1-4]. For instance, in order to obtain an improvement in Epworth sleepiness scale at least 4 h/night of CPAP is required [2], 6 h/night is required for multiple sleep latency test and memory[1-2] , 6h/night is required to decrease cardiovascular risk[5] and 7.5 h/night for functional outcome associated with a sleepiness questionnaire[2]
- What is the clinical relevance for paediatrics?

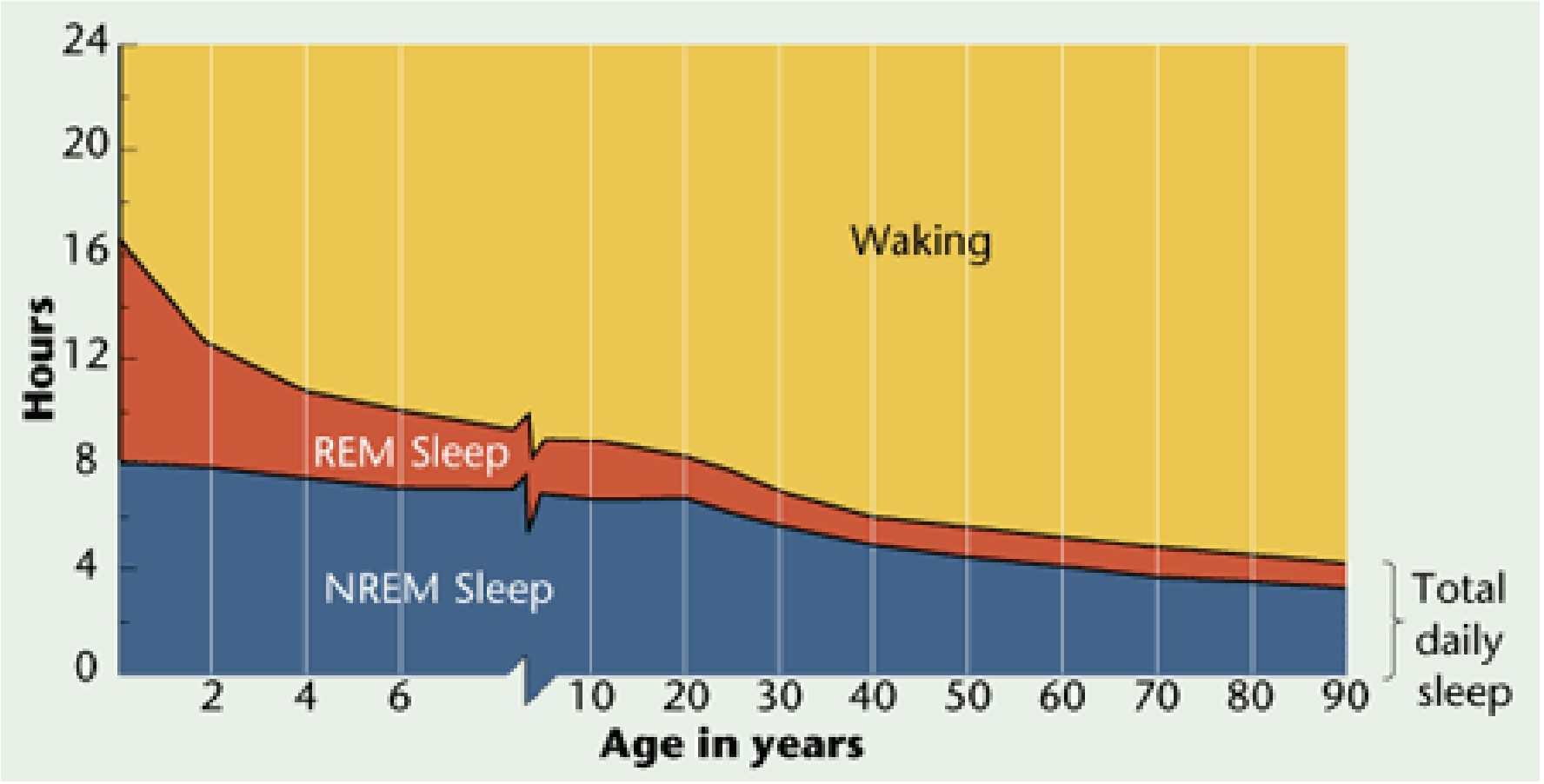
1. *Zimmerman ME et al. Chest 2006 ; 130:1772-8*
2. *Weaver TE et al. Sleep 2007 ; 30:711-9*
3. *Antic NA et al. Sleep 2011 ; 34:111-119*
4. *Stradling JR et al. Sleep 2000 ; 23 Suppl. 4:S150–S153*
5. *Bouloukaki I et al. Eur Respir J 2014 ; 44:1262-74*

Total sleep duration - change with age



Modified from FERBER R.: Solve your child's sleep problems. New York. Simon & Shuster, 1985, p19

Ontogeny of REM and NREM sleep



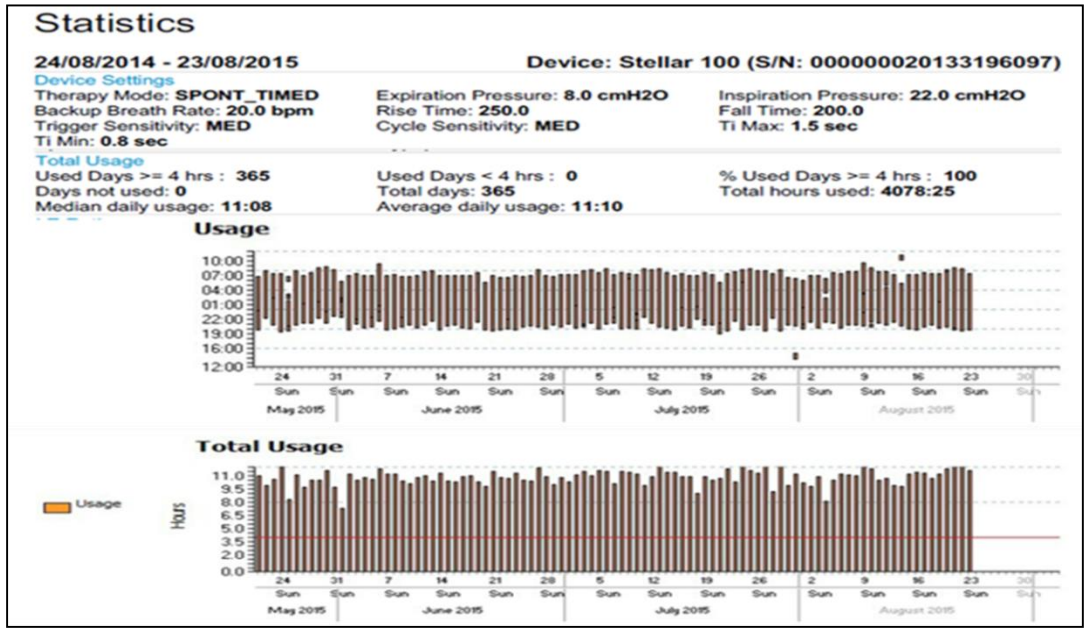
Adherence to NIV

- To date, literature on adherence to NIV in children is limited
- NIV use for OSA amongst adolescents [1]:
  - Higher users had improved attention, grades, school related QoL
  - Lower users more likely to show decline in above
- CPAP treatment for OSA amongst 2-16 yr old[2]:
  - Correlation between adherence and Epworth Sleepiness Scale
  - Improvement seen in behavioural problems and attention deficit
  - Dose –response relationship was not demonstrated
  - Mean use of NIV = 2.8 hours /night!

1. *Beebe DW et al. PLoS One 2011;6.e16924*
2. *Marcus CL et al . AJCCM. 2012; 185:998-1003*

Adherence to NIV

- Discrepancy is often found between subjective reports and objective data on adherence.
- Technologies such as built-in software enabling recording and download of ventilator parameters, usage and events are increasingly used for monitoring of the ventilators and their usage.



Adherence to NIV

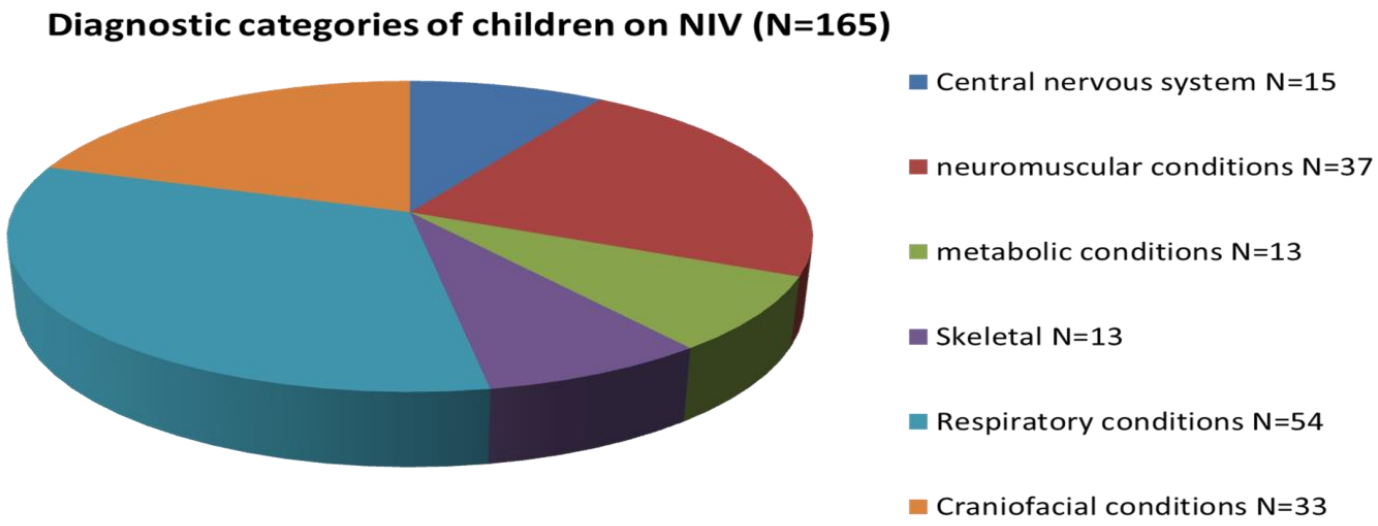
Positive influence	Negative influence
Positive initial encounter	*Technological and equipment issues*
Child and care giver engagement	*African American ethnicity*
Ongoing health education for child and caregiver	*Older age of children (adolescents)*
Peer support group	*Lack of social support*
Individualised strategies for adherence	*Negative feelings towards the chronic illness*
Behaviour modification and rewards	*Not using NIV when away from home*
High maternal education level	*Low maternal education level*
Subjective symptom improvement	*Lack of subjective symptom improvement*
	Fear and embarrassment regarding treatment

1. *Simon SL et al. Sleep Med 2012; 13: 172-77*
2. *King MS et al. Sleep Med Clin 2014;9: 219-234*
3. *Prashad PS et al. J Clin Sleep Med 2013;9: 1303-1313*
4. *Ennis J et al. J Clin Sleep Med 2015;11(12):1409-16*

Adherence in GOSH Cohort
(May 2013- Jan 2015)

219 children on NIV (114 males; age range: 0.6 -20.1 yrs)

- Available data on 165 children:
  - median age [IQR]:11.8 [8.2 – 16.2] years)
  - 93 CPAP; 72 BiPAP users
  - median duration of NIV treatment [IQR] 3.1 [2.0-4.9] years.



Adherence of GOSH cohort

- **Median percentage (%) of days used** was 86.3 %
- **Median usage** 8.3 hours/night.
- **Percentage of days used:**
  - 63% of patients (104/165) achieved ≥ 70% of days used
  - 55% of patients (91/165) achieved ≥ 80% of days used
- **Median hours used/night:**
  - 74.7% of patients (121/162) used NIV ≥ 6 hours/night
  - 55.6% of patients (90/162) used NIV ≥ 8 hours/night

Adherence of GOSH cohort

- **Univariate analysis shows:**
  - ***Age of initiation*** (p=0.03) was significantly associated with % of days used.
  - ***Younger age*** (p=0.01) and ***age of initiation*** (p=0.01), use of ***BiPAP v CPAP*** (p=0.03) were significantly associated with increased median hours used/night.
  - **Gender, diagnostic categories and duration of treatment** were ***not*** related to either marker of adherence (% daily use, median hours used/night)
- **Multivariate analysis** suggests
  - **younger age of initiation** (p=0.005) and use of **BiPAP v CPAP** (p=0.01) remain significantly associated with increased **median hours used/night**.

Laverty A et al. ERS 2015

Adherence in NIV

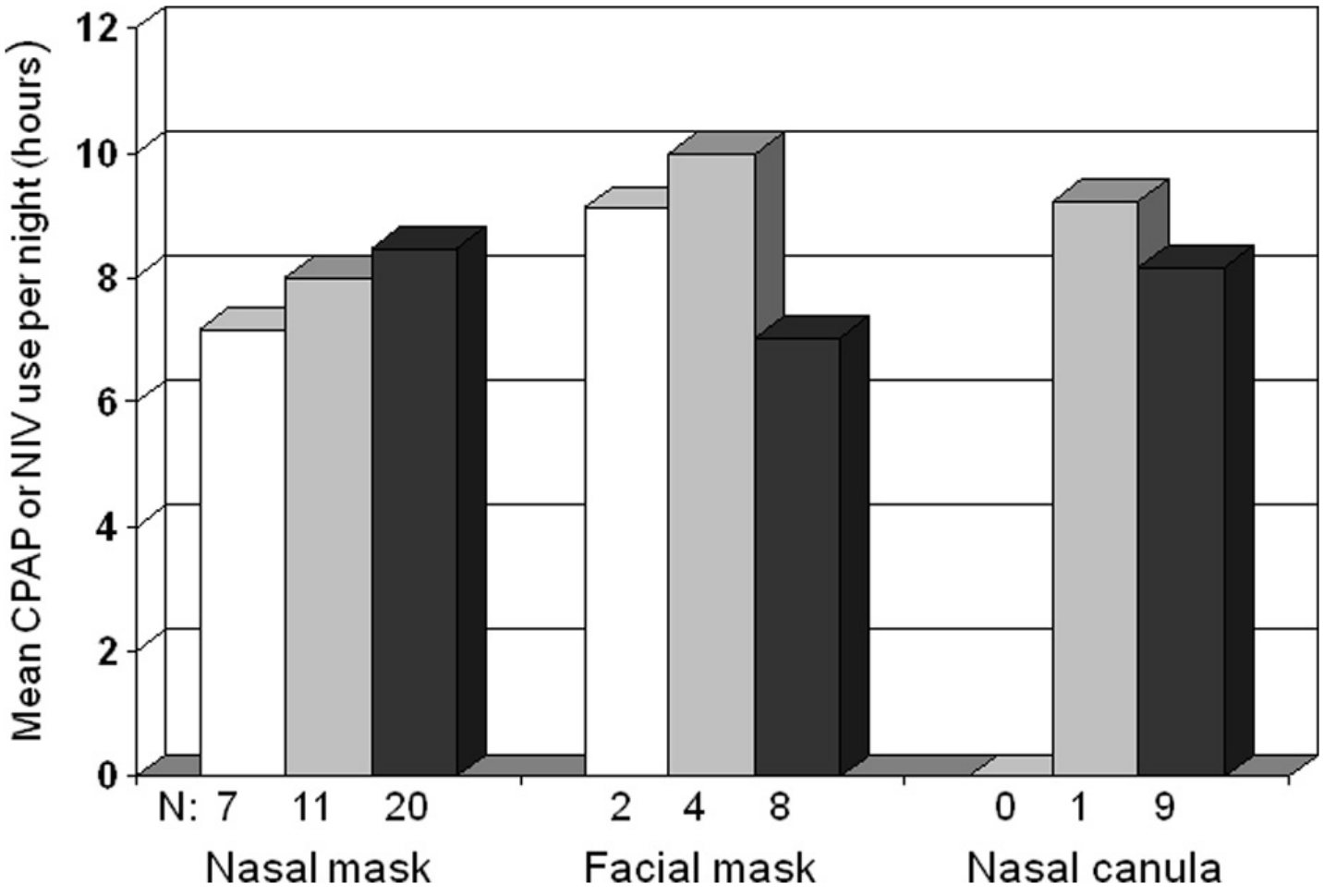
Adherence data reported from other centres:

- Ramirez et al[1] showed:
  - a high adherence (i.e. 72% using NIV >8 hours/night; 86% daily use) among their paediatric patients (total N=62; 51 on CPAP)
  - no difference in adherence between CPAP or BiPAP users.
- Other researchers [2,3] have reported much more disappointing adherence figures, e.g.
  - DiFeo et al: N=56; mean nightly use of 3 hours in the first month
  - Simon et al: N=51; 41% of nights were >4 hours usage

1. *Ramirez a et al Sleep Medicine (2013) Dec 14(12):1290-4*
2 *DiFeo N et al. J Clin Sleep Med (2012); 8:279-86*
3 *Simon SL et al. Sleep Med 2012;13:172-177*

Continuous positive airway pressure and non-invasive ventilation adherence in children

Ramirez a et al Sleep Medicine (2013) Dec 14(12):1290-4



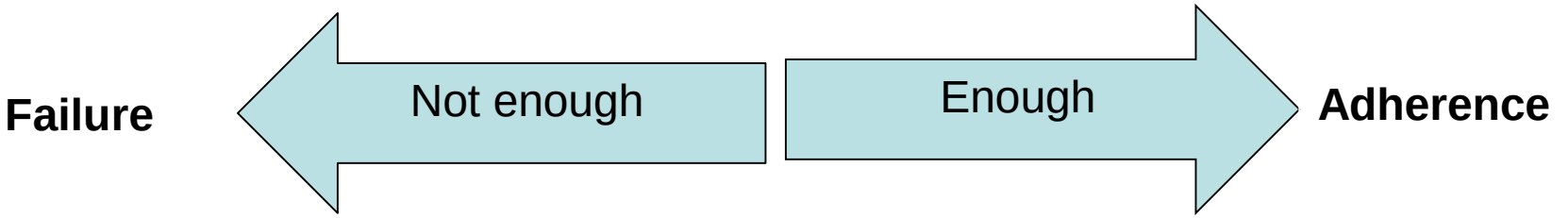
Continuous positive airway pressure (CPAP) or non-invasive ventilation (NIV) adherence assessed as the mean daily adherence in patients according to the type of interface and the duration of treatment. The white bars represent the patients ventilated for less than 3 months; the grey bars represent those ventilated between 3 and 12 months; and the black bars represent those ventilated for longer than 12 months. Treatment duration and the type of interface did not affect CPAP or NIV adherence. The numbers on the x axis represent the number of patients

CPAP/NIV initiation procedure

- Started for all patients with the child's parents in dedicated paediatric NIV unit with well-trained and experienced staff
- Patient discharged home after tolerating at least 6 hours per night after normalisation of gas exchange
- Home visit by home care provider on day of discharge, at 1 week and then every 1-3 months as well as ad hoc if required
- Parents were instructed to contact the NIV unit in case of any problem with the interface or the ventilator
- A systematic sleep study with CPAP or NIV treatment and recording of overnight SpO2 and PtcCO2 was performed in the hospital 1 month after the start of treatment and then every 2–6 months according to the age and the pathology of the child

Ramirez a et al Sleep Medicine (2013) Dec 14(12):1290-4

Adherence and failure are opposite ends of a continuum



Where is the line drawn between non-adherence and failure?

Categories of failures

The failed cases are classified according to main contributing factor to failure:

- **Technical** = inability to apply the mask for a reasonable period of time during sleep because of patient intolerance; inability to tolerate the introduction of positive pressure with a mask in place
- **Clinical** = inability to correct gas exchange towards normal using a non-invasive interface and titrated pressures – either continuous or bilevel
- **Domestic** = failure to provide on-going non invasive support in the home setting despite a clinical success in the sleep laboratory
- **Behavioural/psychological**

Failures in NIV 2008-2015

34/390 patients initiated on NIV failed (25 CPAP; 9 BiPAP):

- technical (n=13)
- clinical (n=3)
- domestic (n=12)
- psychosocial/behavioural (n=2)
- other (n=4)

- the failure rate/year ranged from 4 - 17.9%.
- Overall failure rate = **8.7%.**
- Our data showed that most failures occurred within the first 3 months of initiation.
- In a review of paediatric home ventilatory support in Auckland over a 12 - year period (NIV patients =155; TrLTV= 5), institution of support failed in 11% [1].

1. Edwards et al, J Paediatr Child Health 2005 Dec;41 (12):652-8

How do we maximise the chances of ***success***

Clinical challenge

- Exclude dual pathology
- Consider a bronchoscopy
- Ensure surgical remedial pathologies are addressed
- Consider BiPAP if CPAP requirements are too high
- Range of masks and machines

Technical challenge (the machine)

Potential reasons for non-adherence/failures

CPAP	• Sensation of suffocation • Insufficient pressure
BiBAP	• Poor synchronisation • Autocycling • Poor trigger sensitivity • Rise time insufficient to meet ventilation needs
Both	Pressure sore Dry mouth/nose Inability to communicate Leaks

Technical challenge (the process)

Diagnostic study

↓

Acclimatisation

- Meeting play therapist and NIV CNS
- Mask fitting
- Shown ventilator
- Suggest exercise at home to desensitise wearing the mask

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Elective admission for NIV trial (2 night)

- Titration
- Training of parents
- Issue of equipment

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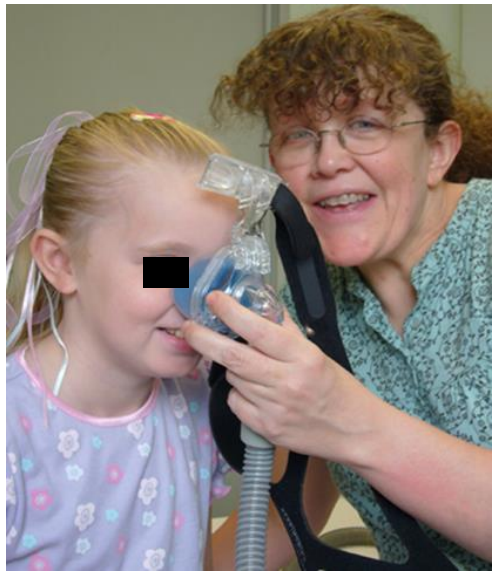
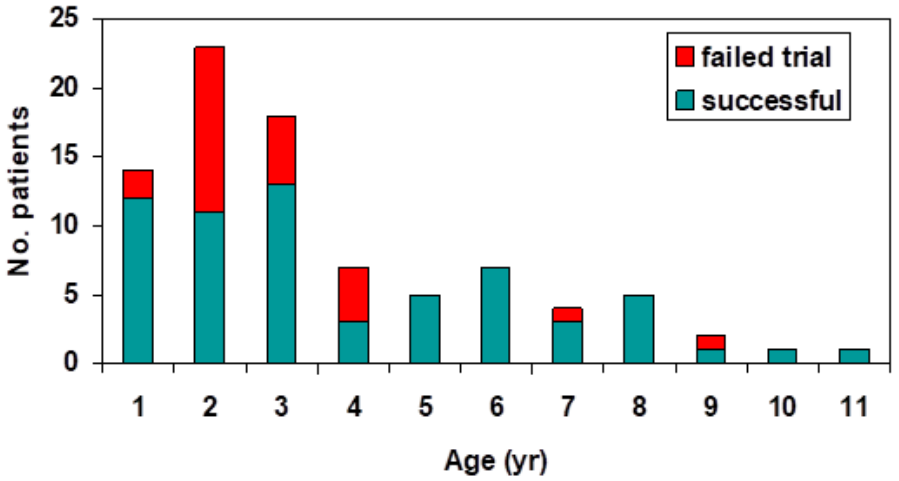
Telephone support
Follow up NIV clinic appts
Follow up sleep studies





Domestic challenge

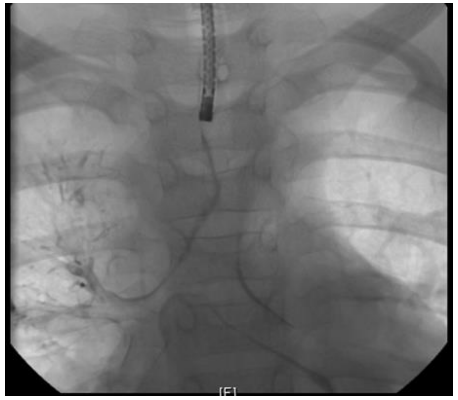
- **support staff** *involved early on* (e.g. allied health specialties such as psychology, play specialists)
- **Home support services** ensures continuous support for patients/families
  - Telephone advice service
  - close links with community teams
- **Follow up clinics**
- ***The family must be on board***

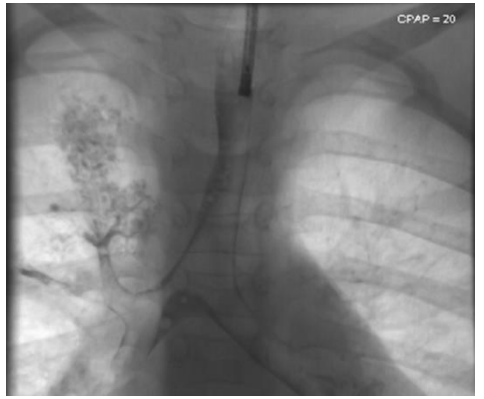
Scenarios that are associated with increased chances of non-adherence / failures…

- Required >16 hours a day
  - unless palliative
- The small and very weak child
  - SMA 1
- Severe craniofacial narrowing
- Behavioural disorders
  - Down syndrome; The terrible two's
- Marked sleep disruption
  - eg some cerebral palsy
- Uncontrolled oral secretions
  - bulbar palsy
- High risk of vomiting / aspiration
- Failure to correct gas exchange
  - not necessarily aiming for perfection

What do we do with total failures?

- Upper airway review
- Tracheostomy
- Nasopharyngeal prong
- Oxygen
- Tackle psychosocial or domestic issues
- Opportunities to revisit NIV
- Do nothing





Concluding remarks –NIV adherence

- Commonly held belief that adherence affect efficacy of NIV
- Huge importance of having support staff and time when providing a NIV service
- Working together: the family must be on board
- Recognise that there are a few situations where NIV cannot be achieved in children
- Unanswered questions about adherence:
  - Yet to establish a clinically meaningful definition of adherence in children
  - Minimal duration of its use to achieve optimal effectiveness remains unclear

Concluding remarks – NIV adherence

- Factors influencing adherence vary across cultures, ages, underlying conditions and practices – this needs to be taken into account when clarifying what these factors are
- Limited longitudinal data to address difference in adherence between different cohorts
- With the right resource, moderately high adherence can be achieved particularly terms of % daily use and hours used/night.
- The younger the age of initiation, the more likely the child is adherent with NIV (both in % daily use and hours used).

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Research project:
Adherence to long term NIV in children

1. Identify barriers and facilitators to adherence in NIV in children by interviewing all major stakeholders
2. Identify what are relevant clinical outcomes and how they are affected by adherence

Thank you for you attention

Any questions?

francois.abel@gosh.nhs.uk

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