

Multi-disciplinary Home Ventilation Program for Patients with Neuromuscular Diseases in Queen Elizabeth Hospital

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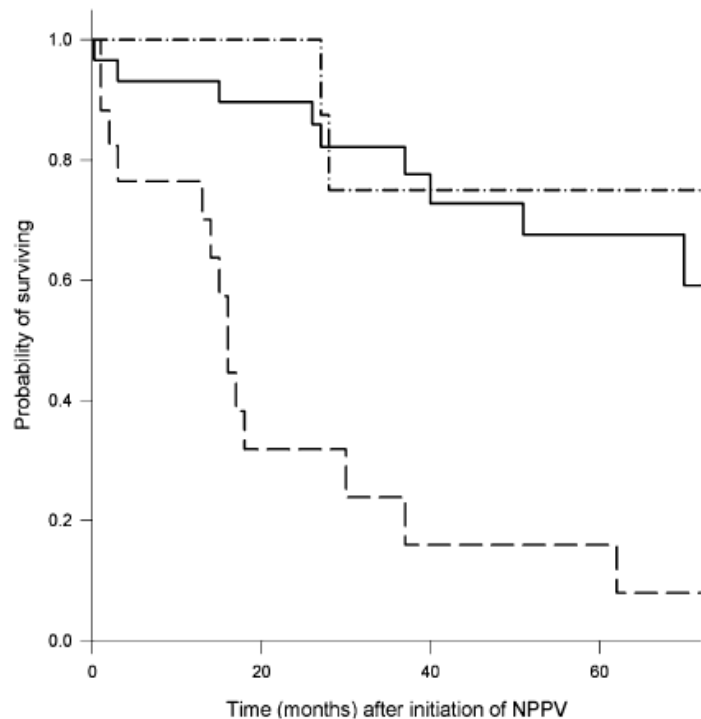


Figure 1 Kaplan–Meier survival curves in patients with (—) slowly progressive neuromuscular disorders, (---) amyotrophic lateral sclerosis and (-·-·-) Duchenne muscular dystrophy. NPPV, Non-invasive positive pressure ventilation.

Shneerson JM. Eur Respir J 2002; 20: 480–487

Neuromuscular Diseases (NMD)

Rapidly progressive

- Motor neuron disease/ALS

Variable progression

- DMD
- Limb girdle MD
- Myopathies
 - Nemaline
 - Metabolic
- Merosin negative congenital dystrophy

Slowly progressive or non-progressive

- Previous poliomyelitis
- Facio-scapulo-humeral MD
- Type III SMA
- Central hypoventilation
- Spinal cord injury

Pulmonary complications and respiratory failure account for 84% of death

Patient with NMD developed acute on chronic respiratory failure and hospitalized



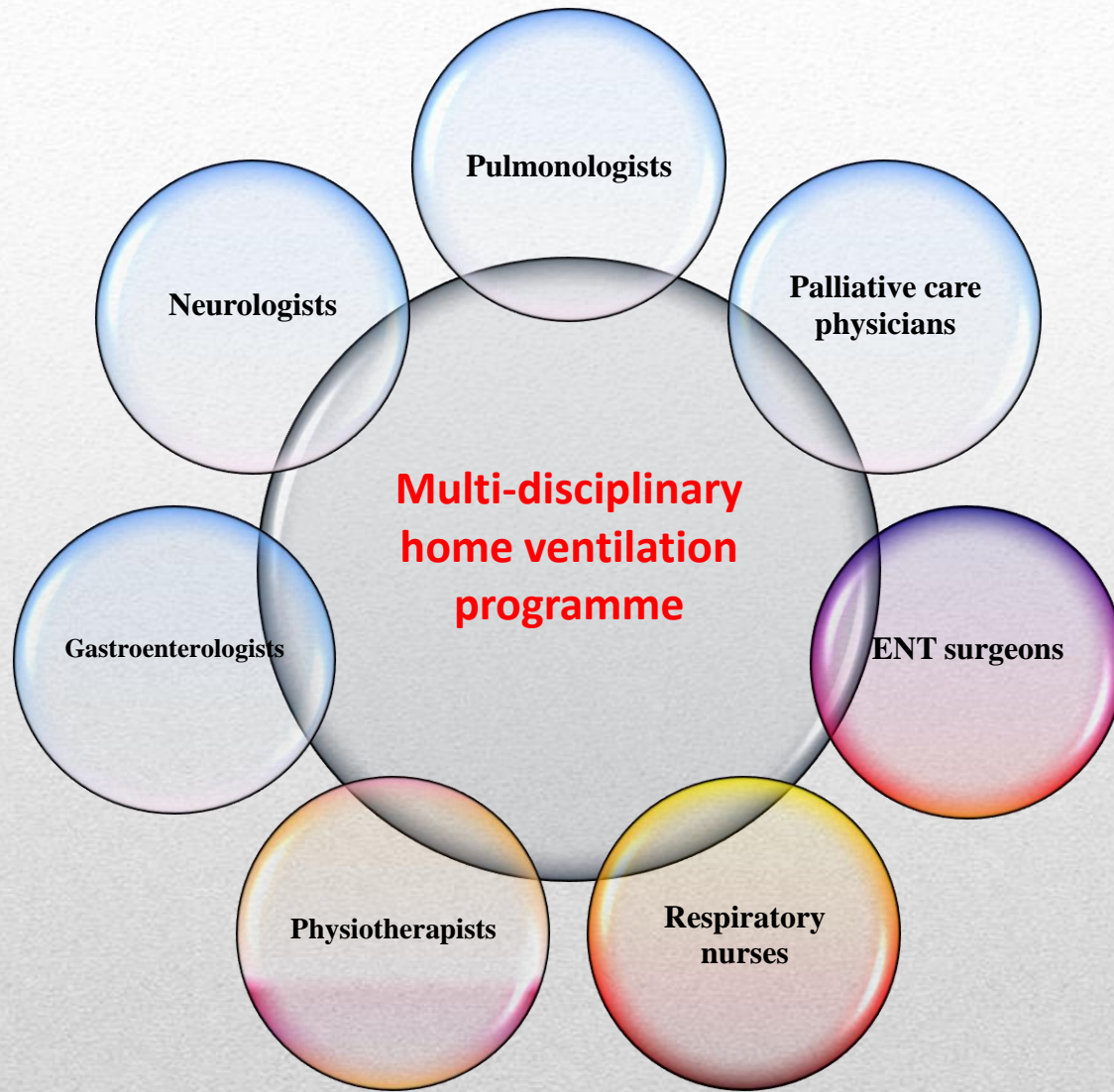
- Unplanned intubation and ventilation
- Prolonged hospital stay
- Excessive death



- Different expectations between relatives and staff
- Conflicts between relatives and staff

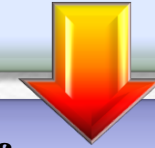


- Long term ventilation plan
- Priming and palliative care plan



Service Improvement Programme

Reduce unplanned intubations and mechanical ventilations



Assist and prepare patients and carers to prepare for HMV before onset of respiratory failure



Regular and comprehensive monitoring of disease progressions

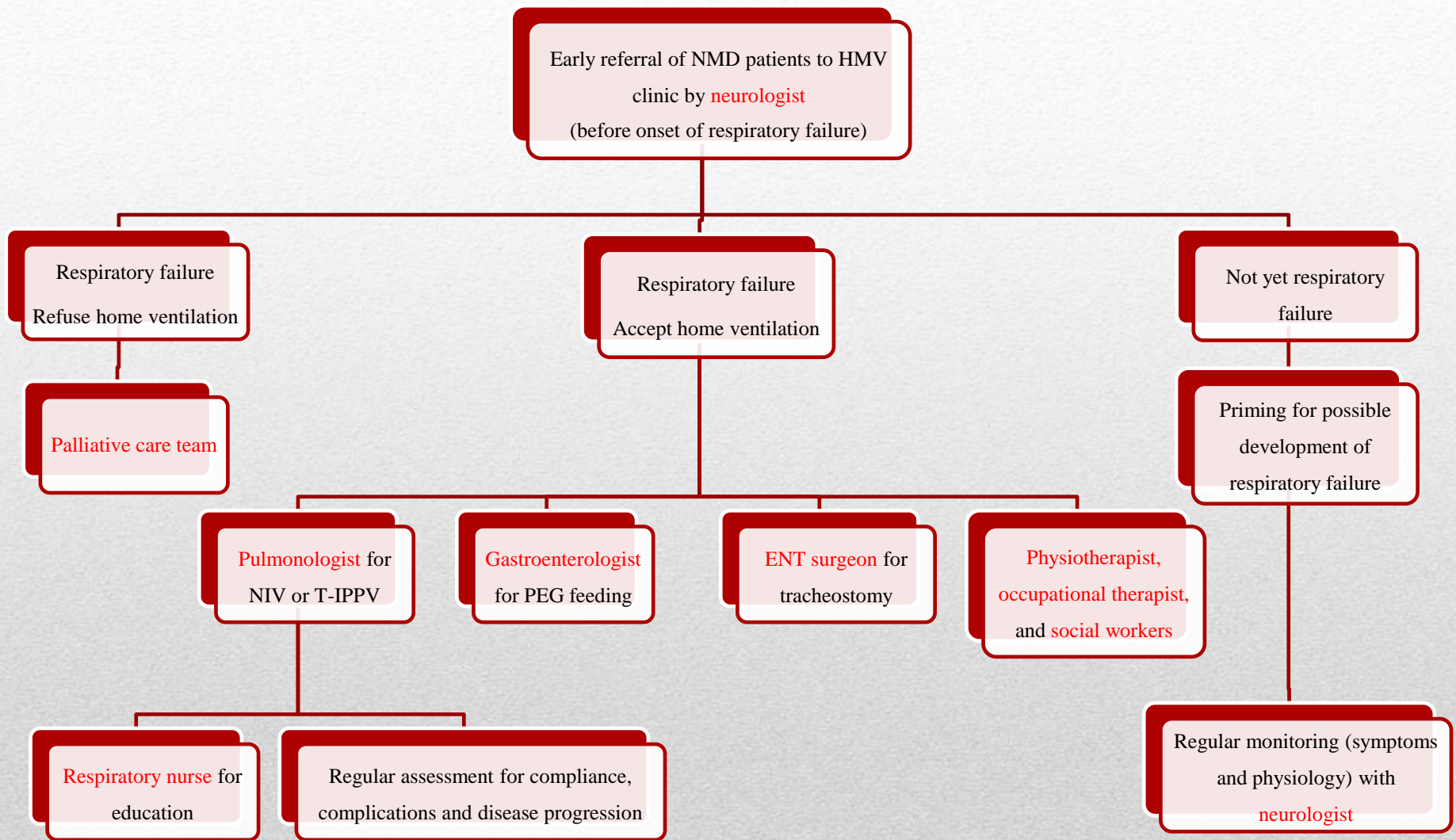


Regular assessment of complications associated and compliance to ventilation



Provide advance care planning and palliative care if necessary

Deliverables

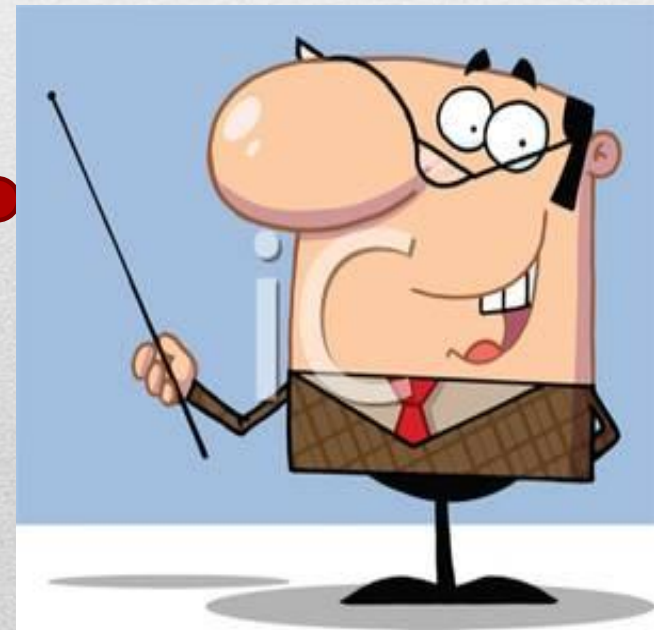


Multi-disciplinary Home Ventilation Service Model

Audit Methodology

All consecutive patients that were recruited prospectively to the program since 2007 were included

Data were retrieved from hospital records and analyzed with SPSS software



39 NMD patients attended the HMV programme since service establishment

Baseline Demographics	NMD Patients (n=39)
Age [years]	56.13 ± 16.59
Male Sex [number (%)]	24 (61.5)
Smoking history [number(%)]	8 (20.5)
Underlying NMD [number %]	
• MND	28 (71.8)
• DMD/SMA	3 (7.7)
• Critical illness neuropathy	2 (5.1)
• Others	6 (15.4)
Bulbar symptoms [number (%)]	17 (43.6)
Riluzole tried [number (%)]	10 (25.6)
Co-morbidities [number (%)]	16 (41.0)

Unplanned intubations and early use of NIV

Unplanned intubation and ventilation

- Ventilated before HMV clinic assessment = 12/12 (100%)
- Ventilated despite HMV clinic assessment = 4/8 (50%) (p= 0.014)

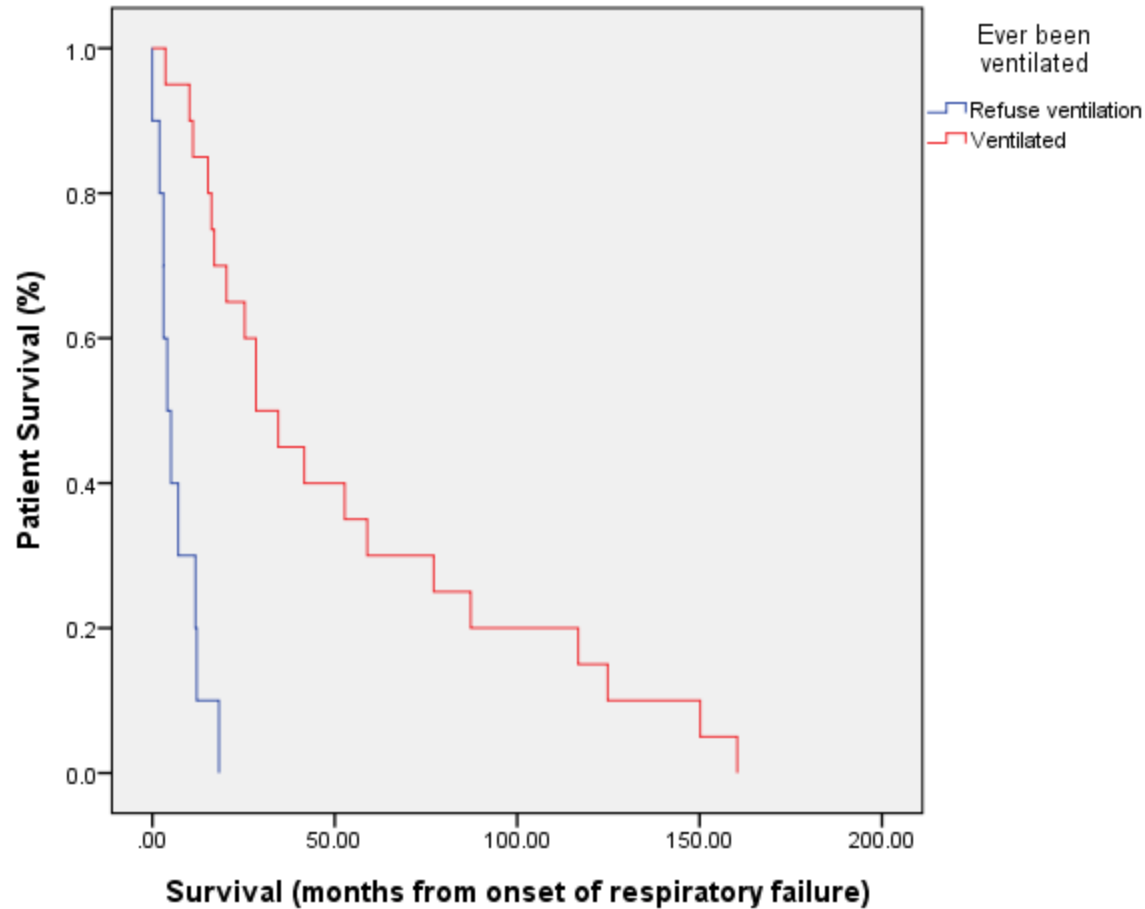
Early use of non-invasive ventilation

- Before HMV clinic assessment = 4/12 (33.3%)
- After HMV clinic assessment = 6/8 (75.0%) (p= 0.17)

Mortality

	Number of patients
Crude mortality rate [number (%)]	21 (53.8%)
Causes of mortality [number (%)]	
• Respiratory failure ± pneumonia	13 (61.9)
• Aspiration pneumonia	3 (14.3)
• Upper GI tract bleeding	2 (9.4)
• Acute myocardial infarction	1 (4.8)
• Stroke	1 (4.8)
• Intestinal obstruction	1 (4.8)
Disease-specific mortality [number (%)]	
• MND	19 (90.5)
• Critical illness neuropathy	2 (9.5)
• DMD/SMA	0 (0)

Survival in Ventilated Patients



	Ventilated group n= 20	Non ventilated group n= 10	Difference (Log rank test)
Medians survival (months)	31.5 (IQR 16.4-84.7)	4.6 (IQR 2.8- 11.9)	0.005

Complications



Complications [Number (%)]	Frequency
	n=20
Mask interface	4 (20)
Pressure sores	5 (25)
Dry mouth	2 (10)
Machine breakdown	0(0)
Tracheostomy related	5 (25)

- Complications associated with HMV were usually minor and related mostly to face masks or tracheostomy wound
-

BiPAP Compliance Analysis

Compliance Summary

Comment

Close monitor of compliance

Date	17/07/2011 - 23/07/2011
Usage	7 days
Days with device used	3 days (42.9%)
Days without device used	4 days (57.1%)
Cumulative Usage	20 hr 5 min
Max Usage (1 day)	7 hr 54 min
Average Usage (all days)	2 hr 52 min
Average Usage (days used)	6 hr 41 min
Minimum Usage (1 day)	5 hr 39 min
Artefact	9 hr 19 min
Percentage of days with usage >= 4	42.9 %
Percentage of days with usage < 4	57.1 %

	Max	Average	Median
Oxygen Purity			
Oxygen Flow Rate			
Pressure IPAP:EPAP	17.1 : 4.8	16.9 : 4.8	17.0 : 4.8
Respiratory Rate (BPM)	40.0	21.7	14.8
Tidal Volume (ml)	1000.0	700.9	668.8
Flow Rate (l/min)	100.0	75.7	76.2
Leakage (l/min)	72.8	43.6	42.4

Detection of mask leak

95th Percentile Pressure IPAP:EPAP	17.0 4.8 cmH2O
Time Spent with Leakage > mask compensation	0 hr 1 min (0.1 %)

Early detection of disease progression

	Max	Average	Median	5th / 95th Percentile
SpO2 (%)	99.0	93.0	93.3	86.0 / 98.0

Search: [] Details **Alert**

Reminder List Delete

All Date Range
 Current Hospital Only Current User Group Only

Hosp	Creation Date	By	User Group
QEH	24/03/2010	NCK627	MED

Detail New Save

Effective from: 24/03/2010 Period: 9999 month(s) No Expiry

To: 24/06/2843 **FU appointment on 29/07/2011**

Content:

Chronic type II respiratory failure put on NIV since 2/2010

IPAP 15, EPAP 5, RR 10, FiO2 1L/min

Patient opt for NO intubation and invasive ventilation if NIV support failed

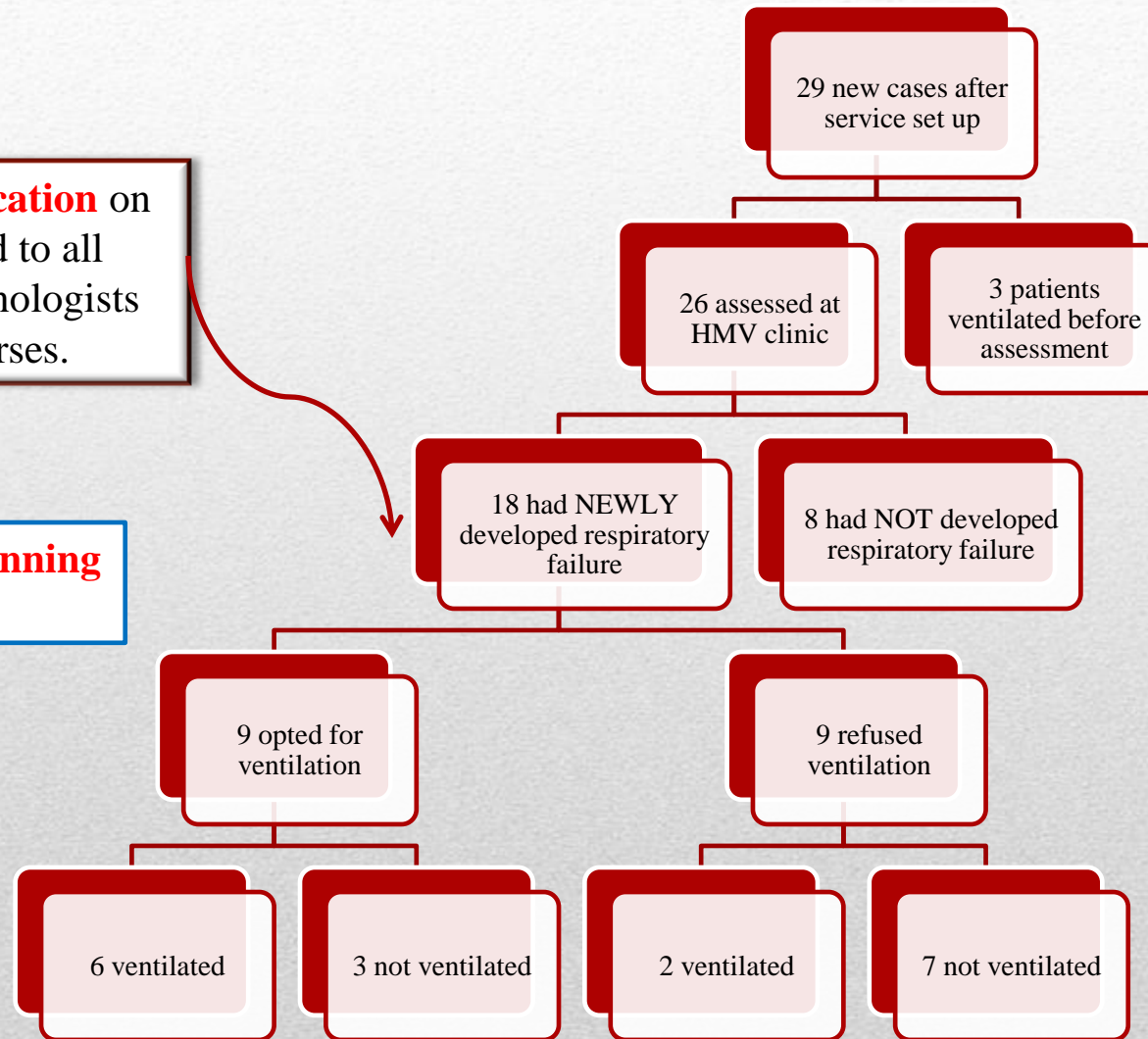
Since 23/5/11, BIPAP setting changed with IPAP: 17, EPAP:5, RR:12, O2: 1.5L

Created by: NCK627 on 24/03/2010 at QEH

Updated by: NCK627 on 20/07/2011 at QEH

Alert on advanced care plans
 agreed ventilation care plan in case of deterioratn are listed for easy reference

Priming and Palliative Care



Priming and education on HMV were offered to all patients by pulmonologists and respiratory nurses.

Compliance to planning
= 13/18 (72.2%)

Feeding through percutaneous endoscopic gastrostomy

- To prevent aspiration and provide nutritional support
- **PEG** feeding was successfully initiated in 14(70%) of the ventilated patients by gastroenterologist.



Conclusions

1

- Home ventilation service significantly reduced unplanned intubations and invasive mechanical ventilation

2

- Improved survival for those with respiratory failure on ventilation support

3

- Patients were primarily discharged to home with non-invasive ventilation (NIV) or continuous positive airway pressure (CPAP)

4

- Optimize feeding

5

- Enhanced monitoring and management of respiratory failure and disease progression

6

- Provision of palliative care

Key to success is

**Multi-disciplinary
engagement and
seamless
collaboration**



Acknowledgement



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