Neurology and/or Palliative Care Consultant Clinic, Physiotherapy
- assess capacity, cognitive, bulbar and UL function
- after diagnosis, when monitoring respiratory function (every 2-3 months), on deterioration or if person asks
- offer to discuss NIV: would patient consider for symptom management and/or prolongation of life
- MNDA NIV leaflet (8A,8B)

Patient wishes active intervention

YES

Assess SpO2 (at rest/on air), FVC and/or SNIP, and symptoms (ensure suitable interface if bulbar involvement); symptoms and signs of respiratory impairment (see NICE 2016 1.14.7) or via local respiratory screening (physiotherapy or respiratory physiology)

All patients with lung disease: (Sp O2 < 92%)

Referral to local respiratory consultant:
Dr Tim Shaw - East Dorset
Dr McConnell - West Dorset

Full discussion re NIV with pt include benefits, disease progression and withdrawal.
Check understanding, d/w MDT (if necessary) families and carers. Consider anticipatory medicines (MNDA Just in Case kit)

YES

NO

With no lung disease:
Sp O2 < 94% or
no symptoms: FVC < 50% predicted or SNIP < 40 cm H2O
with symptoms: FVC < 80% or SNIP < 65 cm H2O men / 55 cm H2O women or rate of decrease >10cm H2O per 3 months

SpO2 > 94% and
FVC > 80% or
FVC > 50% with no symptoms

Respiratory monitoring
physiotherapy/respiratory physiology at least 2-3 monthly consider referral to The Respiratory Centre for overnight oximetry back to

NO

Symptom management
Consider ADRT, Planning for your future, physio referral, anticipatory medicines (MNDA Just in Case kit)

YES

Referral to The Respiratory Centre (TRC), UHS for out-patient assessment (ABG) and discussions
Urgent (within 1 week) if FVC < 30%, Pa CO2 > 6.0 kPa or symptoms of: orthopnoea, daytime sleepiness, morning headache

NIV provided by The Respiratory Centre, UHS (risk assessment, care plan), inform GP, anticipatory meds

NIV

clinical reviews by TRC at UHS in liaison with care by local palliative care/respiratory team/physiotherapy back to

ADRT, consider withdrawal if appropriate

This pathway has been drawn up with all commissioners and service providers involved.