Process to considering institution of non-invasive ventilation

Lead Consultant to have preliminary discussions with family using the 'Factors to considering institution of NIV'

Refer to tertiary Respiratory team

Information gathering
Broad professionals MDT (virtual)
Possible contributors include:
- Lead medical team
- Specialist nursing team
- School nurse
- Education
- Community nursing team
- Respite/hospice services
- Dietician
- Physiotherapy
- Respiratory team

Information gathering
Consider whether any investigations are needed – or whether a pragmatic approach based on clinical assessment is sufficient (esp. if Palliation will likely be the primary management)

Sleep study
Can provide baseline information
Is there a need for respiratory support?
Is NIV potentially able to help?

Family meeting with Respiratory team (and others as necessary)
Collaborative decision making process
Use 'Factors to considering institution of NIV' in documentation
Aims of meeting:
- Detailed explanation of what NIV would involve
- Include burden of care, potential increase as well as decrease in symptoms
- Exploration with family of their views on NIV
- Explaining the need for NIV being an indicator of baseline deterioration and fragility
- Clarifying process to trial NIV
    - What will success look like?
    - What will failure look like? What will be the indicators to stop the trial?
      (When is it too burdensome, uncomfortable, unsafe with secretions or vomiting?
      When there are not the expected gains in reduced hospital/critical care admission?)
- Advance care planning being integral to the process
- The need for ongoing review of the appropriateness of continued NIV
- What is the parallel plan when NIV is no longer appropriate?

Following three actions in parallel

Trial of NIV
Complete Consent form
Trial of mask at home
Admission for 2 nights for NIV

Advance care planning
This should be completed by the team knowing the child and family best
Discussion with the family of their wishes
Consideration of the current level of fragility
Do they have wishes to try and achieve?
What treatments would be in the best interest in the event of deterioration?
Do they have priorities for End of life care?

Palliative care
Support may be useful to all families, but referral should definitely be considered if NIV not used or stopped
What support are the family receiving?
- In care needs?
- In psychological support?
- Is more needed?

Consider CHC or hospice referral
What is the rate of deterioration?
Is support needed for symptom control?

Review
In most children deterioration will occur over time
Regular review of the appropriateness of continued NIV is necessary
Complete "Factors to considering institution of NIV' in review documentation
Regular review of the ACP
Escalation plans may need to change in line with clinical condition
Priorities, wishes and need for support may also change

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Factors to considering institution of non-invasive ventilation?

Patient sticker

NIV is a treatment with potential benefits and harms and the decision process involves necessary balance. [4] This checklist is to provide pointers for clinicians considering whether NIV may be appropriate for a patient and can provide a basis for discussions with a child and family. (This is not to generate a 'score' but to guide clinicians to the relevant considerations, and provide a means for documentation)

- 'Best interests are not just medical best interests. You must consider the person's welfare in the widest possible sense, and consider the individual's broader wishes and feelings, values and beliefs.
- All decisions should follow careful consideration of the individual circumstances of the person and focus on reaching the decision that is right for that person – not what is best for those around them, or what the "reasonable person" would want.
- Those involved in caring for the person, or interested in his or her welfare, must be consulted about their views on the person's best interests, and the person's past and present wishes, feelings, beliefs and values.'[1]

Present for discussion:	**Date of discussion:**
Consideration	**Notes**
What is quality of life like now? Will the addition of NIV bring hoped for benefits? Neuromuscular conditions such as DMD or SMA type 2, or conditions where intellectual impairment is not a marked feature, are likely to benefit from a reduction in symptoms associated with hypoventilation[2]. If interaction is poor with only short periods of wakefulness, then potential improvements are likely to be marginal. If there is intolerance of procedures, then NIV is unlikely to be well tolerated. • What is the child's level of interaction? • Is the child able to participate in activities? Does the child attend school? • Does the child have good periods of wakefulness to benefit from NIV use? • Is there already a significant symptom burden with discomfort or pain?	
How is current physical well-being? Is NIV likely to improve the quality of life? **NIV will add to the burden of treatment.** Severe musculoskeletal constraints require high NIV pressures which are hard to deliver consistently. Nutritional status is an important factor in maintaining skin integrity and tolerance of NIV. Safety in feed delivery is important. NIV should only be routinely administered overnight to allow interaction, opportunity to enjoy experiences and pressure relief. Repeated mask removal for secretion management is burdensome and impractical. The addition of medications during the day (e.g. sedatives or analgesia) to treat symptoms from NIV application is likely negate any gains from NIV. • Is the degree of mechanical failure (scoliosis, marked weakness, poor effort) severe? • What is the nutritional status; are feeds well tolerated and skin condition good? • Can all nutrition be delivered during the day? Is vomiting a feature? • Is the respiratory failure pattern present throughout the day and night? • Is secretion management problematic? Is it likely to need additional intervention and devices (e.g. cough assist) in the near future (making NIV delivery difficult)?	
How has life changed since a year ago? Is NIV likely to improve the quantity of life? If there are other more urgent determinants of deterioration, use of NIV may not provide significant benefit and only prolong dying. NIV will significantly increase the burden of care. Careful explanation of this is required to prevent feelings of parental guilt. If NIV is chosen, a care package and/or prolonged admission *may* be required and this should be factored in if life expectancy is short • Has physical well-being or quality of life deteriorated significantly this past year? • Are there repeated chest crises? – could NIV reduce the frequency/severity of these? • Are there physical factors in condition, such as a rapidly progressive scoliosis, gut failure or neuro-degeneration which are likely to impact life expectancy quickly? • Is disease progression likely to result in NIV failing to deliver relief in the near future? (NIV for >16 per 24 hours is burdensome, with pressure issues and poor interaction) • Would a referral to palliative care be supportive to the family? • What is the current burden of care?	
Having a goal directed approach to NIV use[3] and planning for the future are key As with all treatments, NIV may be appropriate for a period, but may become less so or unnecessary in the future, and this should also form part of discussions. Periodic re-evaluation of the burdens and benefits aids this process, and can take place after a trial period of NIV. Advance care planning discussions with informed joint decision making with families is an imperative. Documenting their wishes in the event of acute or chronic deterioration is vital to communicate to all teams. • Would more intensive respiratory or critical care support be appropriate? • In what situations would NIV no longer be appropriate?	

References

1: https://www.bma.org.uk/media/1850/bma-best-interests-toolkit-2019.pdf

2: Annane D, Orlikowski D, Chevret S. Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders. Cochrane Database Syst Rev. (2014) 13:CD001941. doi: 10.1002/14651858.CD001941.pub3

3: Krivec U and Caggiano S (2020) Noninvasive Ventilation in Palliative Care and Ethical Dilemma. Frontiers in Pediatr. 8:483. doi: 10.3389/fped.2020.00483

4: Ray S, Brierley J, Bush A, et al. Towards developing an ethical framework for decision making in long-term ventilation in children Arch Dis Child 2018;103:1080–1084.

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Patient sticker

Consent form for trial of non-invasive ventilation

Date of consent:	Present for consent discussions:
Discussion area	**Notes**
Diagnosis Clinical concerns leading to consideration of NIV Description of recent clinical trajectory What is quality of life like now?	
Treatment options (Include non-intervention and consequences of all options) What is the proposed treatment? Who will be Clinicians involved in treatment?	
What is the purpose of starting NIV and the expected benefits? What is the likelihood of success? Is NIV likely to improve quality of life? Is NIV likely to improve quantity of life?	
What are the risks in starting NIV? What is the likelihood of these occurring? Discuss the burden of treatment Discuss the burden of care	
Escalation plans in the event of acute clinical deterioration Has an Advance Care plan been completed and circulated?	
Discussions in the event of chronic clinical deterioration What would be indications that NIV is no longer appropriate?	
Follow-up arrangements Discuss possible need for additional care support Discuss possible need for additional clinical support	

Statement of parent: I agree to the course of treatment described on this form and **I confirm** that I have 'parental responsibility for this child:
Signature: …… Print name:…… Date:……..

Confirmation of consent: (completed by a health professional): I have discussed the treatment with the patient and parent/person with parental responsibility and answered any further questions or concerns:
Signature: …… Print name:…… Date:……..