Q+A from 10th Jan 2023: Clinician focus

Respiratory – Dr Anne-Marie Childs

As an adult on 24/7 ventilation, I get no results from Peak Cough Flow (PCF)

Whilst the benefit of regular PCF and other PFTs (pulmonary function tests) in those on continuous NIV (non-invasive ventilation) is less clear, there may be a role in using PCF and PFTs even in patients on same ventilatory support to determine whether ventilation is optimised and to guide use of specific strategies for cough augmentation and secretion clearance. However specific testing will be guided by patient history and symptoms.

Should the relationship between the Brooke score and the PFT be highlighted more in the guidelines?

Given the challenges of height measurement after loss of ambulation, at a time when there is a greater risk of decline in respiratory function, it makes sense to have a lower threshold for consideration of further assessment on those with reduced upper limb strength (as indicated by Brooke Score > 3) and scoliosis. The challenges of height measurement in DMD were discussed a great deal in the respiratory WG. Given impact of less accurate height measurement on predicted values, it is important to assess relative change in absolute values in individual patients.

In patients approaching loss of ambulation, it is helpful to start doing arm-span measurements to determine how these correlate with measured height. The Brooke Score is also useful as a guide to changes in upper limb function and can highlight the risk of respiratory decline but should be considered in conjunction with respiratory symptoms and changes in an individual's absolute / predicted PFTs.

Use of thresholds

Given the fact that height measurement is so unreliable, we have chosen to highlight 'FVC predicted' thresholds that should prompt consideration for referral to a specialist respiratory centre rather than recommending a specific intervention at a particular threshold. FVC measurements fluctuate depending on a number of variables and form only 'part' of the overall picture. The individual's motor function, stage of disease, use of other treatments, particular symptoms or concerns and other measures of respiratory function will all need to be taken into account when determining optimal respiratory management for an individual patient.

The DMD Care UK guidelines are based on a 'typical' disease course but it is important to highlight that some patients (maybe 15-20%) don't follow the 'rules' and can go into respiratory failure earlier and with a level of respiratory function than is 'better' than threshold of FVC 50%. This is particularly true for patients who are obese. The guidelines emphasise the importance of taking symptoms into account as well as 'the numbers'.

It is important to remember that many patients will reach these thresholds of 'increased risk' at a time when they are transitioning from paediatrics into adult

care and ensuring effective and ongoing respiratory monitoring and management is an important element of 'transition'.

Further research into optimal markers / assessments to identify those who will benefit from ventilatory support and other interventions is needed.

What about sleep studies?

Sleep studies should be delivered and guided by respiratory rather than neuromuscular teams. During the pandemic, there have been more outpatient sleep studies – these can be done using oxycapnography (e.g. TOSCAs) at home to measure oxygen and carbon dioxide levels overnight. However, some groups report that home oxycapnography (TOSCAs) in non-ventilated patients is less reliable, but the technology may improve.

New national guidance on respiratory assessment requires annual training for assessors. How are centres managing this now? We cannot now FVC and PCF in clinics unless our neuromuscular physio is trained and assessed regularly.

This new guidance should now be shared widely to be sure that all practitioners are aware and consider the impact of having 'trained assessors' to undertake PFTs.

We state in the DMD Care UK guidance, that the person doing the assessments should be *appropriately* trained. The working group felt it was important that those undertaking PFTs should be experienced in assessing people with DMD. Awareness of patients' fatigue and ability to tolerate testing and the importance of using an appropriate interface is essential to get the most accurate and representative results from PFTs.