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Paediatric lung function

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Q 1. Could you tell me whether the plethysmograph-measured functional residual capacity reference values measured by HÜLSKAMP *et al*. are still widely accepted?
J. Sulc, Prague, Czech Republic

A HÜLSKAMP *et al*. found significantly lower values for functional residual capacity (FRC) in healthy infants when measured using the "new" Jaeger/Viasys MasterScreen BabyBody infant plethysmograph (FRCp) than had previously been published using older equipment. To my knowledge, there are no larger collections of reference data for FRCp using modern equipment. Therefore, for that particular equipment the reference range (not reference values) suggested by HÜLSKAMP *et al*. is still accepted. However, the authors emphasise that the choice of reference data will always depend on the equipment used, the protocol applied and the population studied. So, ideally, in a research setting it is recommended to concomitantly study an appropriate healthy control group; or at least to check the reference data you want to use in your own laboratory by studying some healthy control subjects.

Q 2. What is the pathophysiological mechanism explaining the worsening of spirometry after bronchodilators in some paediatric patients? We sometimes get a plateau in the flow-volume curve just after the peak, a little bit like the "boot leg appearance" seen in COPD adults.
I. Rochat, Geneva, Switzerland

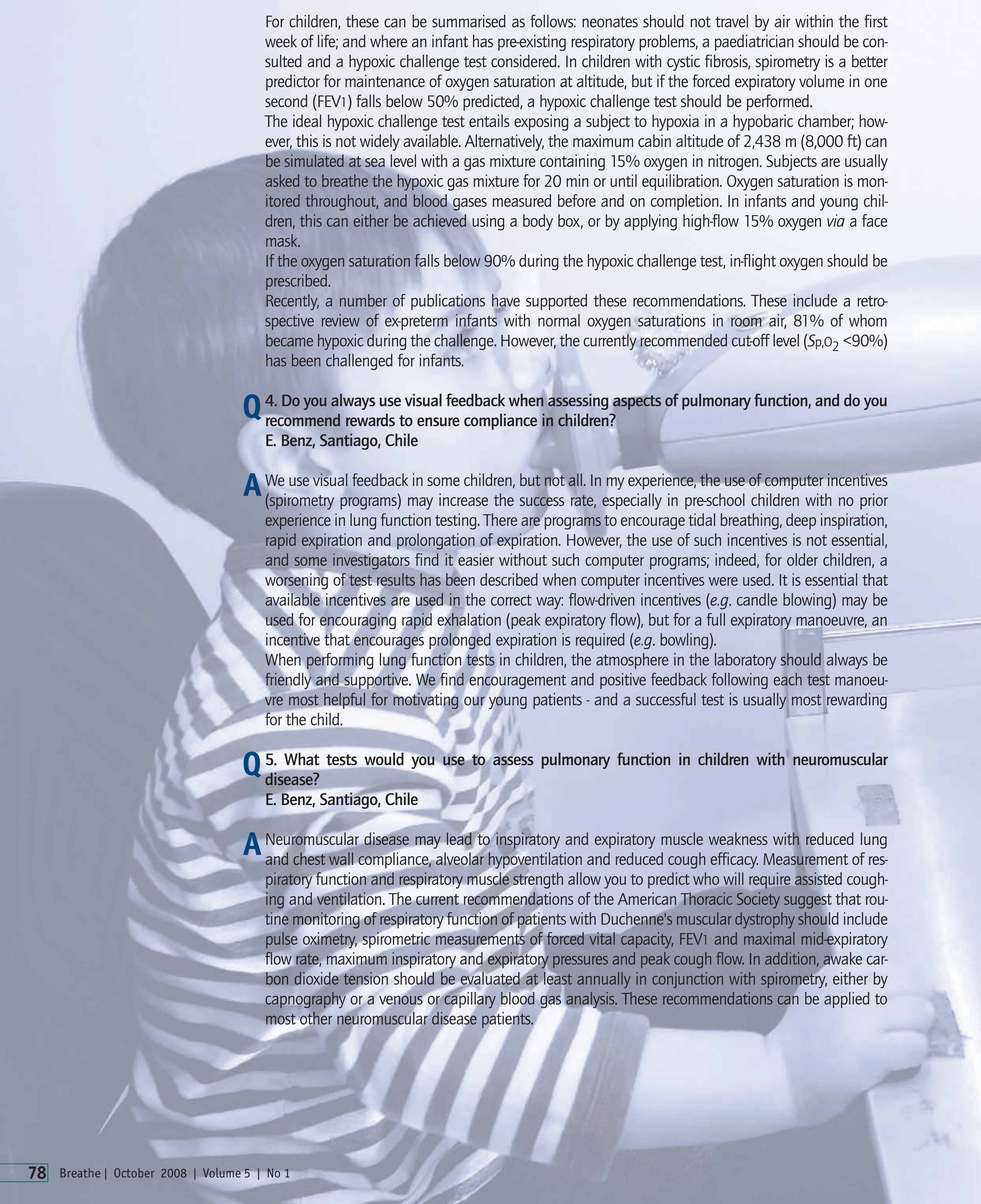
A A paradoxical worsening of the forced expiratory flow-volume curve after administration of a bronchodilator has been described, for instance in patients with cystic fibrosis; this is attributed to increased airway compliance due to loss of muscle tone during a forced expiration. Relaxation of the bronchial wall muscle may result in both an increase in airway diameter and an increase in airway wall compliance. Depending on the anatomy of the individual airway, one of these two mechanisms will dominate the response. If the effect on airway wall compliance is greater with a decrease in the airway diameter during a forced expiration, a worsening of spirometry will result. In children with a chronic obstructive pulmonary disease such as cystic fibrosis you may see a similar appearance of the expiratory flow–volume loop as in adults.

Q 3. Do you carry out 'fitness to fly tests' in children?
E. Benz, Santiago, Chile

A No, currently we do not perform fitness-to-fly-tests in children. However, as the number of people, including children, travelling on commercial flights is increasing, there will be an increasing demand for assessing fitness to fly, which will include children with chronic respiratory disease.

For passenger safety, during commercial flights cabin pressure is maintained at an equivalent of up to 2,440 m altitude. Breathing air at 2,438 m (8,000 ft) is equivalent to breathing 15.1% oxygen at sea level. In healthy subjects, the arterial oxygen saturation as measured by pulse oximetry (S_{p,O_2}) is likely to fall to 85–91% under these conditions, but this is usually not accompanied by symptoms. However, in the presence of chronic lung disease, such as cystic fibrosis or neonatal chronic lung disease, patients may develop hypoxia with respiratory distress, altitude-related illness or even death.

In 2002, the British Thoracic Society published recommendations for patients with lung disease planning air travel. These were updated in 2004.



For children, these can be summarised as follows: neonates should not travel by air within the first week of life; and where an infant has pre-existing respiratory problems, a paediatrician should be consulted and a hypoxic challenge test considered. In children with cystic fibrosis, spirometry is a better predictor for maintenance of oxygen saturation at altitude, but if the forced expiratory volume in one second (FEV_1) falls below 50% predicted, a hypoxic challenge test should be performed.

The ideal hypoxic challenge test entails exposing a subject to hypoxia in a hypobaric chamber; however, this is not widely available. Alternatively, the maximum cabin altitude of 2,438 m (8,000 ft) can be simulated at sea level with a gas mixture containing 15% oxygen in nitrogen. Subjects are usually asked to breathe the hypoxic gas mixture for 20 min or until equilibration. Oxygen saturation is monitored throughout, and blood gases measured before and on completion. In infants and young children, this can either be achieved using a body box, or by applying high-flow 15% oxygen *via* a face mask.

If the oxygen saturation falls below 90% during the hypoxic challenge test, in-flight oxygen should be prescribed.

Recently, a number of publications have supported these recommendations. These include a retrospective review of ex-preterm infants with normal oxygen saturations in room air, 81% of whom became hypoxic during the challenge. However, the currently recommended cut-off level (S_{p,O_2} <90%) has been challenged for infants.

Q 4. Do you always use visual feedback when assessing aspects of pulmonary function, and do you recommend rewards to ensure compliance in children?
E. Benz, Santiago, Chile

A We use visual feedback in some children, but not all. In my experience, the use of computer incentives (spirometry programs) may increase the success rate, especially in pre-school children with no prior experience in lung function testing. There are programs to encourage tidal breathing, deep inspiration, rapid expiration and prolongation of expiration. However, the use of such incentives is not essential, and some investigators find it easier without such computer programs; indeed, for older children, a worsening of test results has been described when computer incentives were used. It is essential that available incentives are used in the correct way: flow-driven incentives (*e.g.* candle blowing) may be used for encouraging rapid exhalation (peak expiratory flow), but for a full expiratory manoeuvre, an incentive that encourages prolonged expiration is required (*e.g.* bowling).

When performing lung function tests in children, the atmosphere in the laboratory should always be friendly and supportive. We find encouragement and positive feedback following each test manoeuvre most helpful for motivating our young patients - and a successful test is usually most rewarding for the child.

Q 5. What tests would you use to assess pulmonary function in children with neuromuscular disease?
E. Benz, Santiago, Chile

A Neuromuscular disease may lead to inspiratory and expiratory muscle weakness with reduced lung and chest wall compliance, alveolar hypoventilation and reduced cough efficacy. Measurement of respiratory function and respiratory muscle strength allow you to predict who will require assisted coughing and ventilation. The current recommendations of the American Thoracic Society suggest that routine monitoring of respiratory function of patients with Duchenne's muscular dystrophy should include pulse oximetry, spirometric measurements of forced vital capacity, FEV_1 and maximal mid-expiratory flow rate, maximum inspiratory and expiratory pressures and peak cough flow. In addition, awake carbon dioxide tension should be evaluated at least annually in conjunction with spirometry, either by capnography or a venous or capillary blood gas analysis. These recommendations can be applied to most other neuromuscular disease patients.

Background picture ©Dr Graham Hall

Q 6. If you were developing a pulmonary function laboratory from scratch to monitor lung function in children predominantly with asthma and cystic fibrosis, what equipment would you view as essential, and what would be ideal?
E. Benz, Santiago, Chile

A Spirometry remains the most commonly used test of lung function in children. Therefore, a spirometer suitable for use in children starting from approximately 4 years of age is essential. You should make sure that the equipment you intend to buy has hardware (*e.g*. handling, mouth piece, dead space, bacterial filter/hygienic aspects) and software (*e.g.* adaptation of scales to paediatric requirements, computer incentives, appropriate reference data) that comply with international standards and are designed for use in children. However, spirometry and forced expiratory manoeuvres will not allow full assessment of respiratory function and therefore will not allow detailed monitoring and differential diagnosis. For a more advanced paediatric lung function testing laboratory, additional essential equipment includes a whole-body plethysmograph and equipment for measuring diffusing capacity. For monitoring and safety, you should have a pulse oximeter available, as well as resuscitation equipment. If your patient population includes a large proportion of pre-school children, equipment to assess respiratory resistance during tidal breathing may be helpful (interrupter technique, forced oscillation technique).

The ideal equipment will always depend on your patient population and whether you intend to set up a laboratory for clinical purposes only, or whether you want to perform lung function testing in research projects. There will be no single answer to the question "Which test is best?" or "Which equioment is best?".

Further reading

Question 1

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