Lung function measurements as a clue to aetiological diagnosis

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Case history

A boy was born prematurely in the 31st week of pregnancy, with a birth weight of 1,470 g and length 42 cm. Due to respiratory complications, he was mechanically ventilated for 5 days and, thereafter, he received continuous positive airways pressure (CPAP) for 2 weeks. He needed oxygen supplementation for 10 weeks.

At the age of 1 yr he developed pneumonia with a major atelectasis and was mechanically ventilated for 1 day. At the age of 1.5 yrs he suffered bronchiolitis due to respiratory syncytial virus positive (RSV+ve) bronchiolitis. Following this, he was treated with regular daily nebulised budesonide (Pulmicort®) and salbutamol (Ventoline®) until 6 yrs of age. Nebulised treatment was then discontinued. Respiratory symptoms reoccurred and he was then started on regular treatment with inhaled fluticasone (Flutide®) and salmeterol (Serevent®) by Discus®.

When the patient was able to perform lung function testing, it was noted that he had a reduction in maximum expiratory flow/volume loops. This did not respond to trials of increased doses of inhaled steroids. He did not experience acute exacerbations of his respiratory symptoms, but he had marked limitation of his ability to take part in physical activity, which was attributed to exercise-induced asthma.

Due to reduced lung function and exercise-related respiratory symptoms, he was referred to Voksentoppen BKL at the age of 10.5 yrs.

Clinical examination

The patient presented as a moderately overweight 10-yr-old boy, with an increased anterioposterior diameter of thorax and breathing with moderately heightened shoulders. On auscultation, he had slightly symmetrically reduction of respiratory sounds in the posterior lower thorax.

Task 1.
What diagnostic steps should be taken?

Answer 1.
As shown below, diagnostic steps that should be taken include: 1. lung function measurements, 2. exercise tests and 3. bronchial responsiveness assessments.

Diagnostic steps

1. Lung function measurements

A maximum expiratory flow/volume loop was obtained for the patient and is shown in figure 1.

Task 2.
How would you interpret the flow/volume loop?

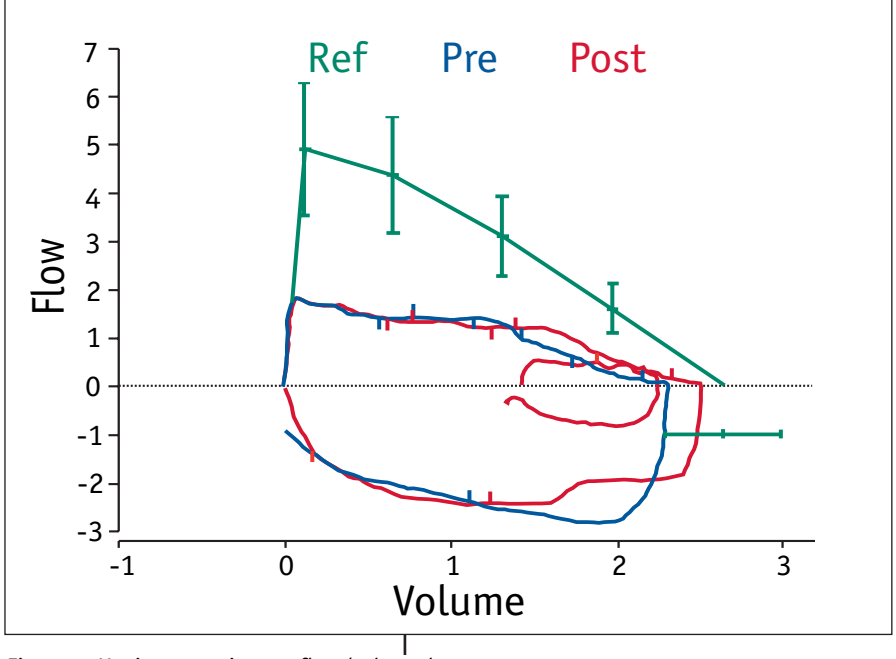

Fig. 1. – Maximum expiratory flow/volume loop.

Answer 2.
The flattened initial part of the maximum expiratory flow/volume loop and markedly reduced peak expiratory flow rate indicate a centrally located airways obstruction. In addition, there was no evidence of reversibility to inhaled salbutamol.

2. Exercise test

There was no reduction in forced expiratory volume in one second (FEV_1) observed after running on a treadmill. Marked audible inspiratory stridor during maximum exercise was noted. The assessment of maximum oxygen uptake was 60% of the predicted value.

3. Bronchial responsiveness measured by metacholine inhalation

The concentration of metacholine which caused a 20% fall in FEV_1 ($PD_{20,met}$) was measured as 0.19 μmol. This corresponds to marked bronchial hyperresponsiveness.

Task 3.
What is your preliminary diagnosis?

Task 4.
What additional diagnostic steps should be taken?

Answer 3.

The lung function measurements and exercise test results suggest a centrally located extra-thoracic bronchial obstruction as the most probable explanation. Bronchial hyperresponsiveness may be related to reactive airways after bronchopulmonary dysplasia in the newborn period.

Answer 4.

As shown below, X-ray of the trachea and HRCT of the thorax, fibreoptic laryngeo tracheoscopy, and CT of the trachea with virtual construction.

Additional diagnostic steps

X-ray of the trachea and HRCT of the thorax

X-ray of the trachea revealed no pathological findings (not shown). A high-solution computed tomographic (HRCT) scan of the thorax revealed minor areas with air trapping and a slight volume reduction in the right lower lobe; however, there changes were very minor and do not explain the symptoms which were observed during exercise (fig. 2).

Fig. 2. – HRCT of the thorax showing air trapping and volume reduction of right lower lobe.

Fibreoptic laryngeo tracheoscopy

Fibreoptic laryngeo tracheoscopy showed signs of laryngomalacia in the supraglottic area (fig. 3) and severe tracheal stenosis with a reduction of tracheal lumen of 50% (fig. 4).

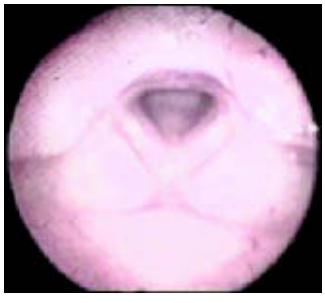

Fig. 3. – Fibreoptic laryngeo tracheoscopy showing minor laryngomalacia in the supraglottic area.

Fig. 4. – Fibreoptic laryngeo tracheoscopy showing severe tracheal stenosis.

CT trachea with virtual reconstruction

Figure 5 shows a virtual "bronchoscopic" reconstruction of CT trachea, which demonstrates a thin web formation in the trachea.

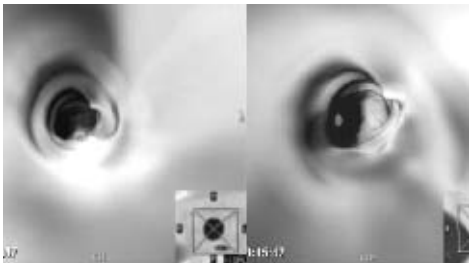

Fig. 5. – Virtual reconstruction of CT trachea showing thin web formation.

Diagnosis:
Tracheal web after tracheal intubation in infancy. Reactive airways disease after bronchopulmonary dysplasia.

Task 5.
What is your suggested treatment?

Answer 5.
Surgical removal by argonlaser of tracheal web and scarring.

Follow-up

After 3 months the patient experienced improved physical capacity with much less exercise limitation, including improved participation in physical activity ("new life").

Lung function measurements were repeated and the results are shown in figure 6; there was marked improvement in the shape of the curve, although not normalisation.

Reversibility to salbutamol was demonstrated; FEV_1 increased 16% (increased up to 86% of predicted value). The curve with measurement after inhaled salbutamol suggests bronchial obstruction, in agreement with the history of bronchopulmonary dysplasia. Tracheo bronchoscopy was repeated and demonstrated minor scarring of no obvious clinical significance.

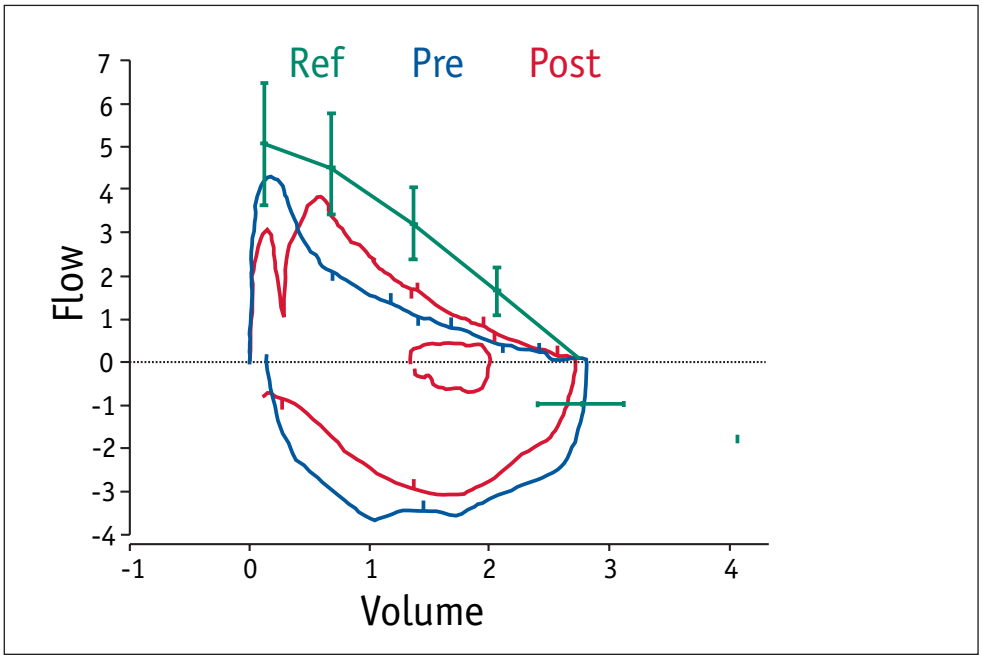

Fig. 6. – Maximum expiratory flow/volume loop at follow-up.