Reviewer's report

Title: Pulmonary Function in Patients with Huntington's Disease

Version: 2 Date: 18 March 2014

Reviewer: Berne Eriksson

Reviewer's report:

Discretionary Revisions

1. In the Introduction (p. 4 line 7 and 8) the authors state that no information on respiratory function in HD is available. There is a study on HD and pulmonary function (A PILOT STUDY ON RESPIRATORY FUNCTION IN PEOPLE WITH HUNTINGTON’S DISEASE Jones et.al. J Neurol Neurosurg Psychiatry 2010 81: A42-A43). This study is only presented as an abstract. However it makes the statement disputable. The authors might consider to correct the writing.

2. A question mark arises on the reference values used (Stanojevic 2009 prediction equations, p. 7). The statement lacks a reference and it is not clear if these equations contain normal values for both FVC and SVC. Further it is not clear why the researcher selected Stanojevic equations and why they are for the population being tested. Since the study is has a case-control design reference values have importance mainly for valuing the pulmonary function in the control group. The reference values also have implications when assessing a restrictive pattern and determining chronic airway obstruction when using LLN (Lower limit of normal) or when determining the degree of obstruction. The authors could consider clarifying this.

3. The authors could make it more explicit that the lung function parameters affected by HD probably only depends on muscle weakness and not to a pulmonary disease since basically only parameters related to forced respiration are affected. Thus a possible pattern of restriction or chronic obstruction really mirrors a neuromuscular disease and probably to a lesser degree lung diseases.

Minor Essential Revisions

1. The aim is clearly defined but in such a general manner (“to compare”) - that the research question (hypothesis) is concealed (p. 4).

2. The authors avoid assessing prevalence of COPD in the groups. This is probably deliberate, but not expressed. Based on the lung function data presented in table 3 it is possible that more subjects in the HD group have a spirometric pattern of COPD with a FEV1/max(FVC SVC) < 0.70. since the spread of SVC is larger in this group and FEV1 lower than in the control group. It is also possible that the HD group show more of restrictive spirometry since both SVC and FVC of normal are obviously low. The variables of lung function in the study make it possible to determine prevalence of spirometric pattern of COPD and its stages and also of restriction. The authors could either comment on omitting these analyses or include them with commentaries.
3. The authors refer to ATS/ERS standards for spirometry but do not include a reference (the reference to ATS/ERS - 14 - is concerned with respiratory muscle testing). The authors do not declare if bronchodilator (reversibility test) was used. The manuscript could include this information.

4. The statement in the Discussion (conclusion) on age (p. 9) is misleading. The control group is age-matched and reference values are age-dependent. Lung function decline is certainly age-dependent. The statement depends on an “in-group” (sub-group) analysis (data not presented in the article). Is it that the authors refer to HD subjects not showing increased ageing in lung function parameters? This writing should be revised.

5 The authors hint at clinical implications of the present study (p. 12 line 8-9 and in the Conclusion section). Early detection of respiratory deficiencies is said to be important to prevent severe respiratory complications. It is of great interest if the authors could be more precise in what ways this could be of value for physicians and for patients with Huntington’s disease.

6. There are previous studies on HD with some similarities. There is a study on muscle weakness in HD showing lower limb strength (published in full) and a study on lung function decline in HD, only in abstract form (Jones 2010). Neither of these are referred to.

**Level of interest:** An article of outstanding merit and interest in its field

**Quality of written English:** Acceptable

**Statistical review:** No, the manuscript does not need to be seen by a statistician.

**Declaration of competing interests:**

I declare that I have no competing interests' below