



Guidelines for clinical pharmacological practices in Huntington's disease.

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Résumé en anglais	<p>OBJECTIVE: Evidence-based medicine is a difficult goal to achieve in rare diseases where randomized controlled trials are lacking. This report provides guidelines that capitalize on both the literature and expertise of the French National Huntington Disease Reference Centre to optimize pharmacological therapeutic interventions for Huntington's disease (HD).</p> <p>MATERIAL AND METHODS: HD experts conducted a systematic analysis of the literature from 1965 to 2013, using a scoring procedure established by the French National Authority for Health. These experts offered their views when evidence was missing to set up provisional guidelines for care in HD. These guidelines were then scored and amended through two subsequent online questionnaires (using SurveyMonkey scoring), and one face-to-face meeting with an external multidisciplinary working group as a step towards validation.</p> <p>RESULTS: Except for the beneficial effects of tetrabenazine in chorea, none of the published recommendations were grounded on established scientific evidence. Second-generation antipsychotics are nevertheless the first choice for patients with psychiatric manifestations (low level of evidence). All other guidelines are based on low-level evidence and little professional agreement.</p> <p>CONCLUSION: Patients' care has greatly improved over the last few years despite the lack of high-level evidence standards. Guidelines are based on the expertise of trained specialists from the French National Plan for Rare Diseases. This strategy should now be extended internationally to promote future studies and to harmonize worldwide care of HD.</p>
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