

10 General Approach to Hyperkinetic Movement Disorders

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Introduction

Movement disorders can be divided into two forms: **hyperkinetic** and **hypokinetic**. Hyperkinetic disorders are also called dyskinesias. There are five forms of dyskinesias which include **tremor**, **myoclonus**, **tics**, **chorea** and **dystonia**. The hypokinetic movement disorders comprise the parkinsonian conditions. There can be rare patients with a mixed movement disorder.

Because a number of different pathologies can cause similar clinical syndromes, a new comer to the movement disorders field can find it difficult and confusing. However, recognition of the different forms of movement disorders with regard the phenomenology is an important first and using a step by step organised way of dealing with a movement disorders patient can lead to rewarding results.

It is important to mention that when faced with a movement disorders patient the basic steps, namely a good history and examination, are vital as one would carry out in any neurological patient. However, there are certain aspects which one does pay extra attention to in a movement disorders case. With regard the history, drug and family history are important points of attention as a number of movement disorders are drug-induced or inherited. With regard the examination special attention is to be paid to eye movements, gait, postural reflexes, and axial rigidity, and tests for bradykinesia as needed as these are not mentioned in the routine neurological examination which often concentrates on the pyramidal system.

General Approach

Three steps are suggested when seeing a movement disorders patient (this exercise was in fact developed in the movement disorders fraternity with the advent of videotapes being made of patients with unusual movement disorders and sent over for an opinion without much history or clinical details).

The **first step** is to distinguish whether the patient has a hyper- or hypokinetic movement disorder. With regard the hyperkinetic movement disorders it is important to try and characterise which variety of the dyskinesia the patient best fits into. Is it a tremor, one of the three forms of jerks (chorea, tics, or myoclonus), or dystonia. Certain features are typical of particular forms of dyskinesias. For example, although both myoclonus and tics are jerky movements, tics are suppressible while myoclonus is not. Also, the patient may describe a rising inner tension when suppressing tics. Ocular jerks are also typical of tics (so-called oculogyric tics). Chorea is typified by jerky movements flitting from one body part to another, in contrast to tics which are stereotyped. Quasi-purposeive movements, where the patient seemingly uses a part of the involuntary movement to do a task (e.g. adjusting the spectacles or the hem of the dress) is also typical of chorea. Dystonia is characterised by abnormal postures, but tremor can be a major feature. The presence of a sensory geste is generally seen in dystonia but not other dyskinesia.

The **second step** is to note the distribution of the movement disorder whether it is focal, multifocal, generalised or unilateral. Is there a particular predilection for a certain body part to be affected or a particular pattern. It is important also to note the age of the patient and whether there are any other neurological features apart from the movement disorder.

The **third step** is to put together the above signs to decide the aetiology and order the necessary investigations. In this regard, when attempting to form a list of possible causes of the movement disorder in a particular patient it is helpful to remember that in general causes of movement disorders fall into four categories:

- 1) **Primary or idiopathic.** These tend to be pure movement disorders (no other neurological or systemic features such as cognitive decline, epilepsy, spasticity, organ failure) which are non-progressive and usually do not have a structural brain lesion. These disorders are often inherited.
- 2) **Secondary.** These movement disorders caused by identifiable secondary causes such as brain injury, infection, or drug use. Other systems may be involved, and other neurological signs may occur. The defining feature of these conditions is the presence of a precipitating factor, and it is important to remember that sometimes a disorder may be delayed after the precipitating event (i.e. tardive movement disorders). Secondary movement disorders are usually static, and do not progress if the precipitating factor is no longer active.
- 3) **Heredo-degenerative.** These are movement disorders which occur as part of a generalised degenerative process affecting the nervous system. Clinical presentation is often variable in these disorders and the movement disorder may only be a minor part of the neurological systemic dysfunction. These disorders have a progressive course.
- 4) **Psychogenic.** This is also an important cause of movement disorder which is a diverse presentation usually coupled with other unusual physical symptoms and signs and often (but not necessarily) the presence of psychological disturbance.

Examples

Applying the three steps, let us look at some examples of how one may apply these steps. Let us take a patient where in Step 1 you decide that the movement disorder is hyperkinetic and it is chorea. In Step 2, you note that the movement disorder is generalised. It is now important to consider the age, the distribution or predilection for a particular area or a particular pattern of the movement disorder. It is also important to look for other abnormal signs. In this patient, apart from chorea, there is nothing else. The patient is 15 years old, so you think of rheumatic chorea, systemic lupus or anti-phospholipid syndrome as the main diagnostic possibilities, if you have excluded drugs. However, if a patient with generalised chorea was 55, rather than 15 years, you would consider Huntington's disease as the most likely diagnosis. So age and distribution were the determining factors in reaching the differential diagnosis. If this patient had a particular pattern of the chorea, for example involving the peri-oral region, then drug-induced tardive dyskinesia would be the first possibility to consider. If the 55 year old had a unilateral chorea or ballism, then a vascular or structural cause would be more likely than Huntington's disease.

Planning investigations

It is important to exclude treatable movement disorders. Conditions such as Wilson's disease or dopa-responsive dystonia are rare, but there should be a high index of suspicion for these.

The differential diagnosis of heredo-degenerative movement disorders is usually very long and investigating the different possibilities is difficult and expensive. In such cases, it is helpful to consider the signs and symptoms carefully to narrow down the differential diagnosis. For example, the differential diagnosis of a patient who has a peripheral neuropathy associated with generalised dystonia is much shorter than of heredo-degenerative dystonia in general. Or if there is deafness associated with dystonia, the list is narrowed to a few causes of heredo-generative dystonias and mitochondrial disease and other disorders which cause deafness and dystonia will be the likely aetiological cause and can be investigated accordingly.

Conclusion

In summary, because of the wide variety of different aetiological causes of movement disorders and a large number of different forms of clinical presentation, it is important to use a step-by-step organised approach to a patient with a movement disorder. The first step is to identify whether it is a hypo- or hyperkinetic movement disorder and if the latter which particular form of dyskinesia. Age and distribution and whether there are associated other signs or clinical features is to be noted. Investigations can be narrowed down by using syndromic associations.

References

Edwards M, Quinn N, Bhatia K. Eds. Approach to Movement disorders. In Parkinson's disease and other movement

disorders, OUP; 2008: 2-7.

Abdo WF, van de Warrenburg BP, Burn DJ, Quinn NP, Bloem BR. The clinical approach to movement disorders. *Nat Rev Neurol.* 2010; 6:29-37.