READING SUB-TEST - PART A

Huntington's disease (HD) | Source texts A-D

TEXT A

Huntington's disease (HD) is progressive. No available treatment has yet been shown to slow, halt or reverse the underlying disease, so management aims to reduce the burden of symptoms, preserve function and optimise quality of life.

The pattern of symptoms changes over time. A medicine that was useful early in the illness may become unnecessary or troublesome later, while a treatment not previously required may become appropriate. The medication list and the reason for each prescription should therefore be reassessed at regular intervals; occasionally, stopping an unhelpful drug is the most useful intervention.

People with HD can be particularly vulnerable to adverse effects of medicines. Prescribers should begin with low doses, increase them gradually and avoid polypharmacy where possible. Many medicines used to control motor or psychiatric symptoms do not have immediate efficacy. Patients and carers should be warned that unwanted effects may be noticed before the intended benefit appears, so treatment should not be judged too quickly unless the adverse effects are severe.

TEXT B

Movement abnormalities are a core feature of Huntington's disease. Chorea is the most recognised motor symptom, but dystonia, bradykinesia, rigidity, myoclonus, tics and tremor may also occur. Bradykinesia refers to difficulty initiating and continuing movement. As control of voluntary movement progressively declines, everyday activities become harder and dependence on others increases.

Early motor changes can be subtle, but in advanced disease almost every movement-related function may be affected. Associated problems can include bowel and bladder incontinence, marked weight loss and pain.

Age at onset influences the initial motor profile. When HD begins in the first decade of life, behavioural or school difficulties may precede seizures, rigidity, bradykinesia and dystonia; chorea is less frequent. Onset during adolescence can resemble the adult form more closely. Adult-onset HD often begins with chorea, while other motor abnormalities may emerge as the disease advances.

TEXT C

Chorea consists of involuntary, irregular, non-repetitive movements of the limbs, face or trunk. The movements may appear purposeless or partly purposeful and are often most prominent in the extremities early in the disease. They are present during waking hours, absent during sleep and cannot be voluntarily suppressed. Stress commonly makes them more pronounced.

Chorea usually increases in severity during the first ten years after disease onset. Later, it may lessen as other movement abnormalities, including bradykinesia, rigidity and dystonia, become more prominent. The pattern is not identical in every patient: some experience a gradual reduction, whereas others continue to have troublesome chorea in advanced disease.

Mild early chorea may be missed during a brief or highly structured examination. Clinicians should therefore observe the patient during spontaneous conversation and movement as well as during formal testing. Walking, turning or performing a second task at the same time may reveal abnormalities that were not initially apparent.

TEXT D

Movement-related risks and management in advanced HD

Motor problem	Likely consequence or risk	Possible management
Chorea	Bruising and abrasions; falling out of bed or a chair; entanglement in long cords	Review medicines; padding of the environment or body; special seating; a floor mattress; remove long cords; monitor for bruises and skin tears
Incoordination of the hands and arms	Difficulty with activities of daily living, including eating	Assistance with daily activities; adapted cutlery and other eating equipment
Gait disturbance	Falls and reduced mobility	Record falls and reassess mobility; use a suitable walking aid; transition to a wheelchair when walking aids are no longer safe or effective; communicate with family
Dystonia or rigidity	Contractures; impaired hygiene; skin breakdown; oral intake may become unsafe or inadequate	Botulinum toxin injections or oral medicines; positioning and skin care; following swallowing and nutrition assessment, consider a gastrostomy tube

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