HUNTINGTON'S DISEASE (HUNTINGTON'S CHOREA) MEDICAL APPENDIX

DEFINITION

1. **Huntington's Disease** is a progressive degenerative neurological disorder clinically manifested by a triad consisting of choreic movements, intellectual decline leading to dementia and emotional disturbances.

CLINICAL MANIFESTATIONS

2. The disease usually begins in adulthood with a peak age of onset of 40 years. It can begin at any age however. It is a rare condition which is relentlessly progressive.

3. Motor symptoms begin with clumsiness and the dropping of objects followed by typical chorea, consisting of brief, low amplitude movements of the fingers. These movements later spread to involve arms, legs, trunk, neck and face. Sustained postures such as a steady grip are difficult and an abnormal gait is characterised by hesitant stuttering steps. Postural instability can lead to frequent falls. Choking is common and sometimes fatal. The disease cannot be diagnosed in the absence of a movement disorder.

4. Emotional disorders are common and may precede choreic movements. Personality changes, mania, hallucinations, delusions, paranoia, hostility and agitation can develop. Depression with apathy and inattention to personal hygiene is common and there is an increased incidence of suicide.

5. Intellectual deterioration appears later and leads to impaired recent memory and judgement. Dementia gives rise to urinary and faecal incontinence and inability to cope with activities of daily living.

AETIOLOGY

6. **Huntington's disease** is a genetic disease transmitted as an autosomal dominant trait. Both sexes are affected equally and each offspring has a 50% chance of becoming affected. A family history is usual but may be concealed by the relatives.

CONCLUSION

7. **Huntington's Disease** is a genetically determined neurological disorder consisting of emotional disturbances, choreic movements and intellectual impairment leading to dementia. External factors do not play any part in the cause or progression of this disease.

REFERENCES

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