Huntington's Chorea
Implications for Nursing in India

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Abstract: **Huntington's Chorea is a rare hereditary diseasec haracterized by chronic progressive chorea and dementia, the exact cause of which is still under study, although many hypotheses are present. The patient has abnormality in gait, choreic movements of the limbs, face, slow mental deterioration and ill sustained depression. At later stages the patient becomes emaciated and succumbs. It is yet untreatable and diagnosed by CT Scans, bio-chemical analysis, family history and classic clinic picture.**

This disease poses a challenge for effective nursing care. There are studies conducted about the nursing aspects—effects of individualized care, effect of musical therapy. It has been shown that diversional therapies, short walks and giving the patient freedom in activities of daily living would help. The prevention at the present day seems to be voluntary sterilization of those at risk after proper counselling.

THERE are many congenital and hereditary conditions that needs a thorough analysis. One among them is the Huntington's Chorea. There is much to be revealed about various aspects of the disease that we have been studying, diagnosing and treating. This is specially true when it comes to the issue of congenital diseases. What is the etiology? What were the pre-disposing factors and so on. Let me put forth the modern issues about Huntington's Chorea.

Definition

It is a rare abnormal hereditary condition characterized by chronic progressive chorea and mental deterioration that terminates in dementia—Mosby's Medical and Nursing Dictionary (1976).

History

It is said that Waters in 1848 in America described this disease for the first time, before George Huntington gave his well known description of it in 1892. He was the third among the family doctors to notice it. His grandfather had recognized it as early as in 1797. But it was presented by Huntington in a paper "Hereditary Chorea", which gave a classic picture of this disorder. He traced this disease in two brothers from Suffolk village who produced 11 generations of choreic sufferers.

Etiology

This genetically transmitted condition is characterized by a gradual change in the sufferers personality as his CNS progressively degenerates. The chorea means dancing and refers to the characteristic writhing movements of the limbs, body and head of the sufferer.

The disease is transmitted by a single autosomal dominant gene and there are 100% possibilities of manifestation of the disease, which was proved by Rosanoff and Handy (1935). However, it is necessary to find out which is the gene that is carried before being manifested. In recent years, it was found that there are some bio-chemical anomalies present. This was proved by Cowie and Seakins (1962), who studied two sons of an affected mother and found them to have raised levels of serum alanine, while the other two siblings had normal values. In another study the affected persons were found to have abnormal levels of serum theta-globulin and gama-globulin fraction.

Studies have been in progress with regard to the etiological factors and we will have to await the results, either to accept or reject the current hypothesis.

Morbid anatomy

Macroscopically there is always an extensive and severe chronic meningitis in which dura may be involved. The brain is atropied, the fronto Rolandic area is most markedly affected. Histologically the small striatal nerve cells are seen degenerating.

The choreo-athctotic movements are thought to be a result of an imbalance of the neuro-transmitters dopamine and GABA (Gama amino buteric acid) i.e., excess of dopamine is present in relation to GABA resulting in chronic excitation of nerve cells which cannot be suppressed because there is nothing to counteract it.

Clinical picture

This disease shows a progressive increase in its clinical features. The early cardinal feature is the suicidal tendency as described by Huntington. Psychiatric anomalies precede the neurological symptomps. In the beginning, the patient may become irritable, moody, ill-tempered and may express the ideas of reference or paranoid delusions. There is increasing slowness and apathy.

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Neurological symptoms like involuntary movements commence in the face, hand and shoulder. It affects the gait of the person. In a study conducted by Koller C. William and Trimble John (1985) on gait assessment, before and after treatment with Haloperidol, 12 of the 13 patients had abnormal gait. The characteristic features included, wide based station, lateral swaying spontaneous knee flexion. There was difficulty in turning, propulsion, which were common. The characteristic feature about this disease, is the way in which, in order to conceal the choreic movements, they are exploited to serve as voluntary actions of quiet unnecessary kind. For example, forward jerk of an upper limb may be converted voluntarily to smooth the hair with hand.

As the disease progresses breathing becomes difficult, thinking muddled, shallow ill-sustained depression leading to suicidal attempts is met with. Memory may be well sustained, although dementia is well established, which is unique in this form of dementiating process.

At later stages, all features increase and patient becomes exhausted and emaciated, bed-ridden, becomes prone to broncho pneumonia and bed-sores. The average duration of illness may be 10-15 years, after which the patient succumbs.

Incidence and risk factor

The average incidence of onset is 30-50 years, though there have been cases of a three-year old developing it. The risk factors at birth are as follows:

*Risk at birth: 50%	After 40 years: 33%
After 10 years: 50%	After 50 years: 12%
After 20 years: 49%	After 60 years: 2%
After 30 years: 44%	After 70 years: 0%

So it appears from the above table that chances of risk reduces as the age increases.

Diagnosis

Although early detection and diagnosis of the disease is difficult, Patterson et al (1948) gave an idea as to how EEG may show on early state of the disease. Recently, Mac Cormac and Michael (1982) conducted a study on attitudes of 40 persons at risk of getting this disease, towards a pre-symptomatic and provocating test, which could possibly diagnose the disease early. They concluded that, persons at risk are receptive for the test, but such testing should not be strongly encouraged.

C.T. Scans can be another diagnostic tool wherein the early change in the structures can be studied. This was proved by O'Donnell B Butters et al (1983) who studied 26 patients and 3 with risk.

Bio-chemical studies may again be useful. But confirmation can be made only after all three cardinal features are present:

1. Positive family history, 2. Involuntary movements and 3. Mental impairment.

Nursing care

Before considering the nursing care aspects of the patients, two studies are presented. (1) Ms Linda, a middle-aged lady who suffered from Huntington's Chorea was in an aggressive and agitated phase. On admission she was assigned to a nurse. The nurse was successful in using nursing process and individualized nursing care. After a lapse of time Linda started recovering to a great extent. Then she regained some memory and even took the nurse to her house. This shows how effective nursing care can be even for the patients with inherited disease like Huntington's Chorea.

(2) Hoskyns Sarah reports that a study was conducted (1982) regarding the music therapy in patients with Huntington's Chorea in which a project was designed where in patients were asked to stay in a holiday home and attend several sessions of music therapy organized by a music therapist. After several sessions, it was concluded that this therapy helps in counteracting the anxiety and frustration, encourages relaxation and also reduction in choreic movements.

It was clear that the therapy served protective, preventive, symptomatic and supportive purposes. Nursing aims at giving the care that embodies the use of all the skills, attitudes and principles in treating the clients with chronic progressively disabling diseases. The nurse controls the environment so that the client does not sustain injuries. Restraints are never used, because of their tendency to increase choreic movements and thus cause injuries.

It would always better to incorporate the nursing process in instituting the nursing care. Planning would help in better patient care. Here, a nurse has to understand, that patient is to be given adequate independence in activities of daily living. With only minimum necessary assistance she should plan for a high calorie and high protein meal, to maintain the body weight and to minimize the risk of decubitus formation.

A nurse should assess the patient's need for diversional therapy and plan accordingly for encouraging and stimulating therapies such as occupational, recreational and physical therapy. Daily walk, light exercises and deep breathing exercises are again helpful. At times it is felt that slower deterioration is seen in patients nursed at home, since they try to cope with family situation and members. The nurse should evaluate her patient for any improvement and change in her plans accordingly.

In terminal stages of illness, nursing care, mental and emotional support is necessary. The family members should be counselled about the patient and how to cope up, by helping him adjust to the family and by assisting him in accomplishing his needs.

Conclusion

As yet there is no available test or predictive test or cure for this disease. Although a genetic marker test is under study from November 1983, it may not be considered sufficiently reliable.

* Small Alwyn, "Huntington's Chorea", *Nursing Times*, April 9-15, 1986, pp. 32-33.

The major problem encountered in counselling the risk families is that they have usually had their children by the time they show the signs of illness. If a person is known to be at risk the problems concerning possible transmission of the illness must be made clear so that they can decide on their having children. So with all the problems, only means of prevention of this disorder appears to be (at the present day) voluntary sterilization of those at risk. Thus it poses a challenge for the doctors and the nurses to treat and manage the patients with this disease.

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Commonwealth Nurses Federation:

The Commonwealth Scene

Several disasters have recently hit a number of Commonwealth countries. In Bangladesh, the floods in September left 3,000 dead and millions homeless. Jamaica was the worst affected Carribean country in the wake of Gilbert in the region's most savage hurricane. In August, an earthquake killed several hundred people in India. In July, the kingdom of Lesotho was cut off by an unusual snow storm. Our thoughts have been very much with our colleagues in these countries, coping with the aftermath of these disasters, and thank them for their devoted work on behalf of the communities they serve.

The Commonwealth Secretariat have now set up a Disaster Management Programme and will commission a series of practical studies to help governments and voluntary agencies to predict and prepare for natural and man-made disasters. Regional nursing networks have been further strengthened by the approval of a College of Nursing for the eleven Commonwealth Countries of East, Central and Southern Africa and the establishment of a South Pacific Nurses Forum. Lord Briggs of Lewes, Chancellor of Britain's Open University, has been elected Chairman of the Governing Board of the "Commonwealth of Learning". The President for the Board is Dr James Maraj of Trinidad, who was a former Assistant General at the Commonwealth Secretariat and Vice-Chancellor of the University of the South Pacific. The Commonwealth Foundation is offering to help meet the cost of providing vidcos on environmental and developmental issues to Commonwealth NGOs in developing countries.

The United Kingdom branches of the Trinidad and Tobago Nurses Association and Nurses Association of Jamaica have celebrated their tenth anniversaries. Throughout this period both organisations have been in regular contact with their colleagues in Trinidad and Tobago and Jamaica to exchange professional information and to support health projects in their home countries. A number of Guyana nurses in the UK have launched "The Association of Guyanese Nurses and Allied Professionals in the United Kingdom," and have elected to support maternal and child health programmes in Guyana. Our congratulations to all concerned for maintaining these vital links between Commonwealth countries.

The Federation's new President is Mrs Margaret Kobue of Botswana. The Vice-President is Mr C. B.P. Kurup of India. Mrs Peggy Vidot of Seychelles is now the elected Board member for East, Central and Southern Africa; Mrs Narender Nagpal of India is Board member for South Asia, and Miss Nadia Osborne of Sierra Leone is Board member for Africa West.