News, Notes and Queries

AN EARLY DESCRIPTION OF HUNTINGTON'S CHOREA

THILENIUS (1816) gave the first description of an inherited form of chorea, and he was followed by Waters (1841), Gorman (1848) and Lyon (1863). In 1872 George Huntington thoroughly described the curious family disease which has since borne his name. Huntington's description, which forms the foundation of our knowledge of this disease, was first presented to the medical world in a lecture at Meigs and Mason Academy, Middleport, Ohio, on 15 February 1872, and was printed in the *Medical and Surgical Reporter* in the same year.

However, an adequate description of inherited St. Vitus's Dance as a disease *sui generis* was given as early as 1859 by Johan Christian Lund, district physician in Saetersdal, Norway, and appeared in his State Medical Report of 1860, a long forgotten document which was rediscovered in 1914 (Hanssen). Lund's report was again referred to in 1938 (Refsum). It is therefore rather surprising to find that Lund's name is not mentioned in monographs on Huntington's chorea.

The relevant passage in Lund's Medical Report of 1860 reads as follows:

> As recorded in the previous medical report, chorea St. Vitus (which is Lund's term for St. Vitus's Dance) seems to recur as an hereditary disease in Saetersdal. It is commonly known as the 'twitches', occasionally as the 'inherited disease'. It usually occurs between the ages of 50 and 60, generally starting with less obvious symptoms, which at times only progress slowly, without becoming violent, so that the patient's normal activities are not particularly hindered: but more often after a few years they increase to a considerable degree, so that any form of work becomes impossible and even eating becomes difficult and circuitous. The entire body, though chiefly the head, arms, and trunk, is in constant jerking and flinging motion, except during sleep, when the patient is usually motionless. A couple of the severely affected patients have during the last days of their lives become *fatui* (i.e. demented). The disease occurs in two families which are registered below. Information is not as complete as could be desired though enough to start with, as long as doctors in Saetersdalen are mindful of the disease in future.

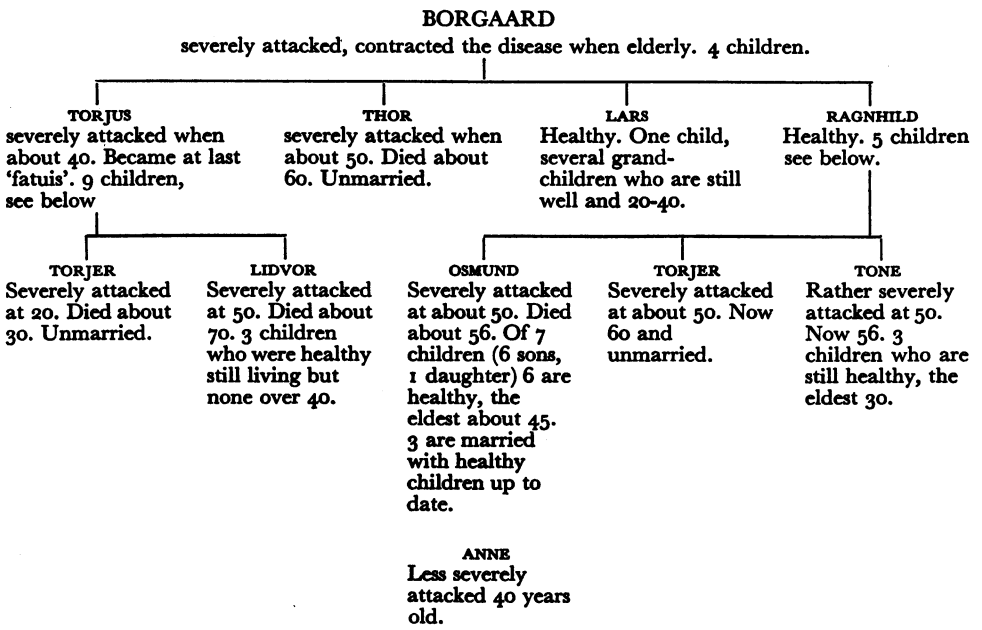
In his medical report of 1868 Lund stated:

> The disease still keeps to the same families and in the same way as reported in 1860. Since then Torgus Lidvorsen (48 years old), son of Lidvor Torjusen from the Valle family has been affected and now has the disease rather badly. In the Byglands family, Mari (Anne's daughter) died some years ago, while her sister Birgith is now severely affected.

Lund's family pedigree through four generations shows that the disease is inherited from one generation to the next and affects both women and men. The course and degree of the disease does not seem to depend on inheritance, as those only slightly affected, or seemingly unaffected, have had severely affected children, and vice versa. In defining the disease according to Lund's description, one would say:

> Chorea St. Vitus's Dance in Saetersdal is an hereditary, chronic and progressive disease which affects adults. It is characterized by choreic movements, and may result in dementia. It affects both sexes.

(VALLE PARISH)



(BYGLANDS PARISH)

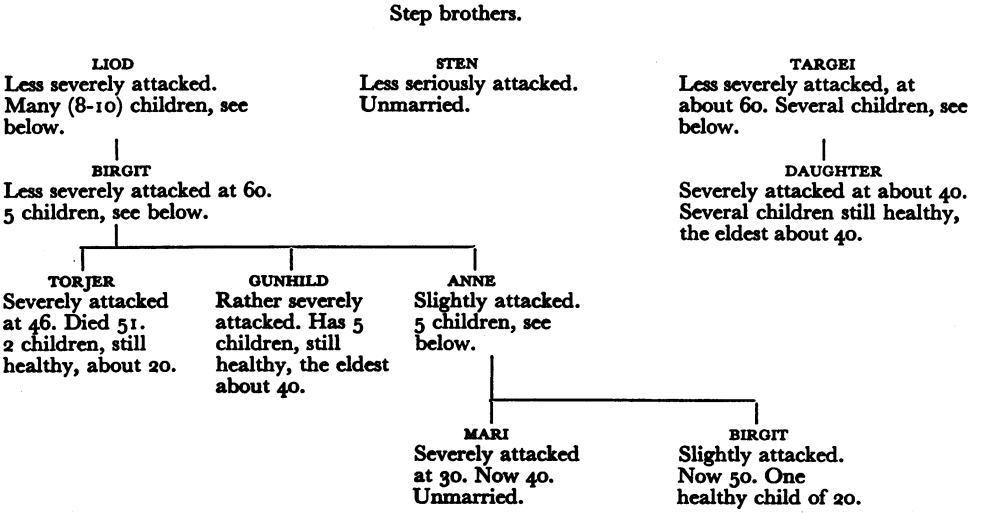


Fig. 1

In 1868 Lund wrote:

In a German journal, the correctness of the diagnosis is questioned. To this I can only reply that I do not have, and never have had, the slightest doubt about it. The picture of the disease when at an advanced stage is as characteristic as possible.

Hirsch, author of the famous *Handbook of Historical and Geographic Pathology* (1862–4), who had read Lund's earlier report, gave the following account of the disease:

One peculiar phenomenon is the endemic prevalence of a form of convulsions in the Säderdal region (Robygeelagets Amt) in Norway, called in the areas concerned 'St. Vitus's chorea', and obviously corresponding to our 'paralysis agitans'. This disease is restricted to particular families but is constantly passed on within these by heredity, and is therefore known among the people concerned as 'Arvesygen' (the inherited disease) or, from the symptoms, as 'Rykka' (i.e. jerks or twitches). The disease usually appears in these people between the ages of 50 and 60, and then with hardly noticeable symptoms which in some cases progress only very slowly and never become very severe, so that those affected are not seriously hindered in their activities. More frequently, however, the symptoms become so severe within the space of a few years that the patient is handicapped in any kind of work and can raise food to his mouth only with difficulty. The whole body, but particularly the head, arms and upper part of the trunk, are in constant movement, with violent jerks or swinging backwards and forwards (flinging, as the account describes it), which ceases only with the onset of sleep; a few of those who are particularly severely affected become demented towards the end of their life. Up to the present there has been no fuller information as to the details.

As will be seen, Hirsch is of the opinion that Lund's chorea (St. Vitus's Dance) is a form of cramp identical with paralysis agitans. The question then arises whether Lund's description could give rise to confusion with paralysis agitans, or on the other hand, that paralysis agitans was so little known at the time as to make possible an erroneous diagnosis.

The symptoms of paralysis agitans were not unknown at that time. James Parkinson had in 1817 described paralysis agitans:

as consisting of involuntary tremulous motion, with lessened muscular power, in parts not in action, and even when supported, with a propensity to bend the trunk forwards, and to pass from a walking to a running pace, the senses and intellect being uninjured.

Lund's description of chorea (St. Vitus's Dance) as a progressive disease where the choreic movements can become excessive, is a sharp contrast to the muscular inactivity of paralysis agitans. The rhythmical tremors in paralysis agitans, with a frequency of four to six a second, are so entirely different from the choreic movements that there is no possibility of confusion. Lund also accentuates the transmissibility of chorea and illustrates this through four generations. He shows the tendency towards dementia while affection of the intellectual sphere is not a noticeable trait in paralysis agitans.

Mutual traits in the diseases, such as the common age of onset and progression, can scarcely be of importance in this connection. On the other hand Lund has not mentioned one single characteristic trait of paralysis agitans in his chorea patients. Lund's chorea therefore has no essential similarity to paralysis agitans, but shows a rather contrasting picture.

The reason why Lund's discovery did not attract attention could certainly not be that his description was so vague as to cause confusion, neither could it be due to the inadequate knowledge of paralysis agitans at that time. The reason rather seems to have been that Lund did not describe the disease in an easily accessible journal.

Furthermore there is the fact that Lund's observations were looked upon as paralysis agitans in one of the largest medical works of the time. Huntington's and Lund's descriptions are similar as to the main points. Both characterize the disease as serious and chronic, a disease which occurs in adults and begins with slight choreic movements progressing in the course of years, until the patient becomes a jerking wreck. They both stress the typical hereditary properties of the disease. Both also mention the tendency towards dementia.

Independent of earlier descriptions, it is now possible, after the lapse of nearly a hundred years, to conclude that Lund's and Huntington's chorea are identical.

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Book Reviews

Sherrington, Physiologist, Philosopher, and Poet. LORD COHEN OF BIRKENHEAD. Liverpool: University Press, 1958; pp. 108. Illustrated. 17*s.* 6*d.*

It is anything but easy to contribute fresh material upon such a theme as Sherrington, who has already been the target of a number of biographers. Lord Cohen has devoted the first of his series of three lectures, delivered at the University of Liverpool, to a straightforward account of Sherrington's career and his achievements in physiology. In Lecture 2, Cohen has sketched the historical background of our present-day knowledge of neuro-physiology, followed by a clear exposition of some of Sherrington's masterly contributions. It is in the third lecture that Cohen breaks new ground by commenting upon Sherrington in his capacity as philosopher and as a poet. Numerous quotations are afforded us, mostly taken from the Rede and from the Gifford lectures, which indicate Sherrington's lifelong preoccupation with the mind-brain mystery. The author concludes that '. . . in this field, Sherrington's thinking-cap was second-hand'—having been already worn by exponents of dualism ever since Descartes. For Sherrington's accomplishments as a poet, Cohen accords high praise. Some element of scepticism must, none the less, be confessed to linger. It is open to serious argument whether Sherrington's poetry was really of a very exalted character, and in our respectful enthusiasm for the master's unique position in science, we should be chary of undue evaluation of his role as a humanist.