Electroencephalography and Clinical Neurophysiology
Elsevier Publishing Company, Amsterdam – Printed in The Netherlands

EEG IN PREDICTION OF HUNTINGTON'S CHOREA. AN EIGHTEEN YEAR FOLLOW-UP

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(Accepted for publication: January 10, 1966)

INTRODUCTION

One of the more tragic features of Huntington's chorea is the delayed appearance of symptoms until affected individuals are well into, or even past, the age of reproductive capacity. It would be desirable for a number of reasons to be able to identify carriers of the gene responsible for this trait before they reach reproductive age.

Patterson *et al.* (1948) examined 26 offspring of diagnosed chorea patients with a battery of tests in an attempt to identify those who would develop the disease in later years EEG examination was felt to have prognostic value Predictions were made which were based on (a) the degree of EEG abnormality, (b) the presence of paroxysmal features, and (c) the presence of slow wave activity over the motor areas.

Twelve offspring with the greatest amount of EEG abnormality were designated "most likely to develop chorea" Seven with lesser amounts of abnormality were given a "bare possibility of developing the disease". For seven offspring with normal EEGs a prediction was made that they "probably would never develop Huntington's chorea".

The purpose of this study is to present a clinical follow-up of predictions for offspring of Huntington's choreics made by EEG 18–20 years ago, and to evaluate those predictions in relation to current neurologic status of those offspring.

MATERIAL

During a statewide clinical and genetic survey of Michigan for Huntington's chorea (Reed and Chandler 1958; Reed and Neel 1959; Chandler *et al* 1960) follow-up clinical information was obtained for most of the 26 subjects in the EEG prediction study. More recently all except two of the offspring were again located. One was excluded from the study since, in retrospect, it seemed probable she already had Huntington's chorea at the time of the prediction.

Eighteen of the remaining 23 have been examined by the author; reliable institutional and medical reports were gathered for the remaining five.

In the family pedigrees which appear in the publication of the original study of Patterson *et al.* (1948), seventeen of the individuals included in the present tabulations may be identified by their generation and individual numbers (Table I)

TABLE I

Kindred No.	Generation No	Individual Nos
261	V	1, 2, 3, 4, 5 and 6
610	IV	10
615	III	1, 2 and 3
672	V	9
756	VI	1, 3, 4 and 5
819	V	2
945	IV	6

The original study included two other kindreds, No. 1073 and No. 1075, but those pedigree charts were omitted from that publication. The five individuals in kindred No. 1073 who also had follow-up studies are in generation VI, individual numbers 10, 11, 12, 14 and 15. Only one member of kindred No. 1075 was included in both studies He is number 21 in generation IV.

RESULTS

Twelve of the offspring eventually developed Huntington's chorea. To date the eleven other subjects have remained clinically unaffected.

TABLE II

The results of an 18 year follow-up on the accuracy of the prediction of the development of Huntington's chorea from EEG studies

Prediction	No. of individuals	Clinical follow-up	
		Choreic	Unaffected
"Most likely to develop" Abnormal EEG	10	5	5
"Bare possibility of developing" Borderline EEG	7	3	4
"Probably would never develop" Normal EEG	6	4	2

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Correlating the twelve diagnosed chorea patients with their EEG prediction (see Table II) it was found that: (1) five were predicted "likely to develop chorea"; (2) four were predicted "probably will never develop chorea", (3) three were predicted as having "a bare possibility of developing chorea".

The three predictions of "bare possibility of developing chorea" were considered incorrect. In those who eventually developed chorea, five had been correctly predicted and seven had been incorrectly predicted

Correlation of eleven unaffected individuals with their predictions are also shown in Table II. Of these five who had been predicted "likely to develop chorea" are now aged respectively 29, 42, 36, 25 and 25 years. The two who were predicted as "will never develop chorea" are now 26 and 38 years old respectively. The four with "a bare possibility to develop chorea" are now aged 27, 33, 45 and 46 years

Assuming that the "bare possibility" group reflects a small chance of developing the disease then six of the eleven in the unaffected group were predicted correctly and five were predicted incorrectly.

SUMMARY

EEG as used by Patterson *et al* (1948) has little value in identifying carriers of the Huntington's chorea trait Clinical follow-up of predictions for 23 offspring of known choreic patients, 18–20 years later reveals 11 were correct and 12 were incorrect

The author wishes to express his appreciation to Dr J. V Neel and Mrs. R Davidson of the Department of Human Genetics of the University of Michigan Medical School and Mrs M. McDonald for assistance in the preparation of this manuscript

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ANALYSE QUANTITATIVE D'ÉLECTROENCÉPHALOGRAMMES RÉPÉTÉS CHEZ LE CHIEN SOUMIS À L'EFFET DU DIÉTHYL LYSERGAMIDE (LSD_{25})[1]

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(Accepted for publication January 26, 1966)

L'étude du LSD_{25} chez l'homme et l'animal en tant que médicament psychomimétique, permettant de reproduire des psychoses aigues expérimentales, a des avantages pratiques, surtout du point de vue de la réversibilité et de la durée plus courte des manifestations psychiques par rapport aux psychoses spontanées chez l'homme.

L'analyse des variations de l'EEG, parmi les autres critères objectifs, se place au premier rang pour évaluer les effets psychomimétiques du LSD_{25} et des médicaments analogues. Sugerman *et al.* (1964) ont montré qu'il y a une similitude étonnante entre l'EEG des sujets traités par LSD_{25} et ceux des schizophrènes chroniques Ellingson (1955), Goldstein (1963) et Goldstein *et al.* (1963b) ont montré que l'étude objective de ces variations n'est pas suffisante et ont proposé d'interpréter l'EEG par les méthodes quantitatives méthode d'intégration automatique décrite par Drohocki (1948) basée sur l'étude de la somme d'énergie des décharges électriques par le cortex cérébral.

Les données de la littérature concernant l'effet du LSD_{25} sur l'activité électrique cérébrale sont nombreuses. Parmi celles-ci on peut relever une série des travaux sur l'effet du LSD_{25} sur l'EEG (Bradley *et al.* 1953; Bradley et Elkes 1957, Bradley 1959, Himwich 1959, Himwich *et al.* 1959, Goldstein *et al* 1963a et b, Hobson 1964), en comparaison avec la schizophrénie chronique (Murphree *et al* 1962, 1964a; Pfeiffer *et al* 1962, Goldstein *et al.* 1963a et b, Sugerman *et al.* 1964), avec les autres médicaments

[1] Le diéthyl lysergamide (Delysid) nous a été gracieusement fourni par la firme Sandoz.