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Computed Tomography for Huntington's Disease

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Summary. CT scans of twelve patients with Huntington's disease were compared with a large series of normal and abnormal scans. The frontal horn/bicaudate ratio in the Huntington's disease group was found to be much smaller than and statistically different from the normal or abnormal CT scan groups. The CT scan appears to be a useful adjunct in the evaluation of patients with suspected Huntington's disease.

Key words: Huntington's disease – Computed tomography – Hereditary chorea.

Until the advent of computed tomography (CT scan), the radiological criteria for the diagnosis of Huntington's disease have been limited to those found on pneumoencephalography [2, 5, 6, 8]. Numerous investigators have found significant relative caudate atrophy on pneumoencephalography using a variety of different measurement indices [2, 6, 8]. These abnormalities, although relatively consistent, are by no means diagnostic. Recent investigations have demonstrated good correlation between measurements made at the time of pneumoencephalography and corresponding values obtained from CT scan [7, 10, 12, 13]. The present study involved identification of the abnormalities on CT scans in patients with Huntington's disease.

Subjects and Methods

Twelve patients with adult onset chorea, family history of chorea and dementia were examined by CT (EMI 80 × 80 matrix). There were nine males and three females, with a mean age of 49.9 years (range 30–80 years) in the study. The average duration of disease was 3.7 years with an average duration of choreiform movements of 3.0 years (Table 1).

CT scans were performed in a standard fashion, at 30° from Reid's line, and the measurements were made from the Polaroid photographs [1, 11]. All measurements were made with a divider, with subsequent recording using a ruler with 0.5 millimeter divisions. Measurements were made in each case using either the 2A or 2B section, whichever demonstrated the shortest bicaudate diameter. A comparison of caudate atrophy to ventricular size was made by calculating the ratio of 2 lines:

1. The maximum bifrontal diameter (FH Line): the transverse distance defined by a line connecting

Table 1. Age, sex, duration of illness, family history, and FH/CC ratios of 12 patients with Huntington's disease

Case number	Age and sex	Years of mental symptoms	Years of movement disorder	Family history	FH/CC
1	48 M	4	3	Yes	1.17
2	48 M	3	3	Yes	1.25
3	52 M	4	4	Yes	1.08
4	32 M	4	4	Yes	1.10
5	60 F	2	1	Yes	1.57
6	40 M	5	5	Yes	1.25
7	56 M	3	2	Yes	1.57
8	80 M	5	5	Yes	1.56
9	44 M	8	5	Yes	1.43
10	55 M	4	3	Yes	1.36
11	30 F	0.5	0.5	Yes	1.29
12	54 F	2	0.5	Yes	1.27

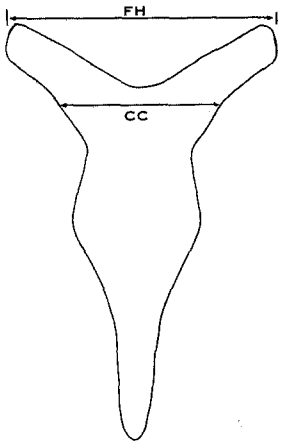
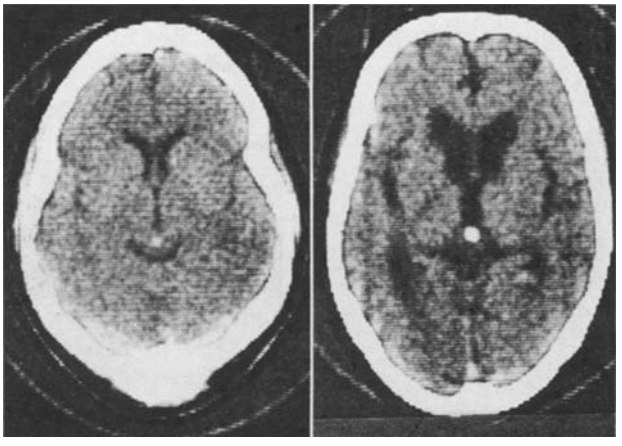


Fig. 1. Schematic representation of lateral and third ventricle with reference to methods of measurement



a b

Fig. 2. **a** Normal CT scan **b** CT scan in a patient with Huntington's disease

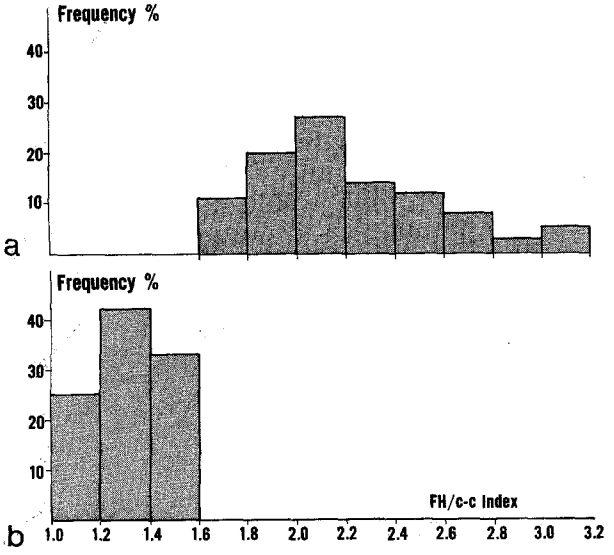


Fig. 3. **a** Distribution of FH/CC ratios in normal-abnormal group **b** in Huntington's disease

the two anterior corners of the frontal horn [13] (Fig. 1).

2. The bicaudate diameter (CC Line): the shortest transverse distance measured between the medial border of the two caudate nuclei (a line drawn between the caudate intercepts of the SC line described by Gath and Vinje [8] (Fig. 1).

A ratio of FH versus CC measurements was calculated for the 12 patients with Huntington's chorea, 26 normal CT scans, and 101 scans fulfilling the CT criteria for cerebral atrophy [12]. The 101 abnormal scans consisted of the following clinical diagnoses: cerebral atrophy 96, Parkinson's disease 5.

Results

The mean ratio of the two measurements in the 12 cases of Huntington's disease was 1.33, with a standard deviation of 0.18 and a range of 1.08–1.57. The mean ratio in the normal group was 2.48 ± 0.35 (range 1.67–3.00) while the mean ratio in the abnormal group was 2.10 ± 0.33 (range 1.6–3.00). The mean ratio of the parkinsonian group was 2.15 ± 0.30 (range 1.72–2.5). The mean ratio of the combined normal-abnormal group was 2.17 ± 0.36 (range 1.60–3.0). The mean ratio for the chorea group is significantly smaller than that for the normal group ($t = 2.92$; $df = 36$; $p < 0.01$), the cerebral atrophy group ($t = 2.04$; $df = 106$; $p < 0.05$), or the parkinsonian group ($t = 2.34$; $df = 15$; $p < 0.05$). There are no significant differences among the means of the non-chorea groups.

The t-test in this case is a very conservative measure of significance since distributions of ratios typically have very large variances. Any non-parametric test would show the difference between the chorea group and the other groups to be highly significant since there is no overlap in their ranges. If we compute a Mann-Whitney U statistic, for example, we find that the probability that the 12 smallest ratios out of 139 all occurred in the chorea group, by chance alone, is approximately 1.4×10^{-19}.

Discussion

Huntington's disease is a generalized degenerative process involving the basal ganglia, cerebral cortex, cerebral white matter, thalamus, and to some extent, the spinal cord [3, 4]. However, the most severely and consistently affected area is the caudate nucleus, particularly the paraventricular portion. There is severe loss of both large and small neurons (especially

small neurons) and marked atrophy and flattening of the caudate ventricular impression on gross examination. There is also cerebral atrophy, but not to such a degree as in the caudate nucleus.

Pneumoencephalography in patients with Huntington's disease has demonstrated good correlation between the radiographic abnormalities and gross neuropathological features. The abnormalities which have been described include:

1) Increased width of the ventricular bodies due to relative caudate atrophy [2, 8, 9].

2) A ratio of the width of the frontal horn to the septum – caudate distance less than or equal to 1.40 [8, 9].

3) Cortical atrophy, especially the frontal areas [2, 8].

However, a normal pneumoencephalogram does not rule out the diagnosis of Huntington's disease [2, 8].

In the present study, CT scans from patients with Huntington's disease were compared with those from normals, cases of cerebral atrophy and Parkinson's disease using the FH/CC ratio. Significant caudate atrophy was found in all cases of Huntington's disease and the findings are similar in pattern to previous pneumoencephalographic work; there appears to be no false positives with various atrophic diseases of the central nervous system using a CT scan FH/CC ratio of less than 1.6 as the criterion (Fig. 3). It will take a larger series of patients to define the accuracy of our measurements, but the present investigation clearly demonstrates the usefulness of CT scanning in patients with Huntington's disease.

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