

DESEZE J., HACHE J.C., VERMERSCH P., ARNDT C.F., MAURAGE C.A., PASQUIER C.A., LAPLANCHE J.L., RUCHOUX M.M., LEYS D., DESTEE A., PETIT H.
Creutzfeldt-Jakob disease: neurophysiologic visual impairments.
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OBJECTIVE:

The predictive value of electrophysiologic visual testing in Creutzfeldt-Jakob disease (CJD) was investigated, and the retinal pathologic findings in three cases are reported.

BACKGROUND:

The fatal prognosis of CJD, its transmissibility, and the lack of treatment make early diagnosis essential in averting human-to-human transmission. Electroretinogram and visual evoked potentials have been studied in few cases of CJD.

METHODS:

A visual electrophysiologic examination was performed in 41 consecutive patients referred with suspected CJD. The disease had been diagnosed in 24 patients (CJD group; 15 were confirmed neuropathologically and 9 by clinicolaboratory methods in accordance with diagnostic criteria). The remaining 17 patients were diagnosed with other neurologic disorders, and served as a control group.

RESULTS:

Flash electroretinogram revealed a significant decrease in the amplitude of the B1 wave (<60 microV) and the B/A ratio (<2) in the CJD group compared with those in the control group. Flash visual evoked potentials revealed no significant difference in latency, but amplitude was increased (>10 microV) in the CJD group, especially in patients with myoclonus.

CONCLUSIONS:

The visual electrophysiologic abnormalities provide an interesting noninvasive diagnostic tool in idiopathic CJD. The B1-wave decrease is closely correlated with the outer plexiform layer abnormalities observed on neuropathologic examination.