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# SUDDEN CORTICAL BLINDNESS FOLLOWING TRANSIENT ENHANCEMENT OF STEROIDS ADMINISTERED IN CONGENITAL ADRENAL HYPERPLASIA

(Case Report)

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A case of cortical blindness developed in an infant who suffered from congenital adrenal hyperplasia and was hospitalized for enteritis is reported. The severe but transient hypertension recorded raises the possibility that the increased doses of steroid administered may have contributed to the pathogenesis of visual impairment.

## INTRODUCTION

Sudden bilateral loss of vision is one of the most frightening events in medicine. In most of the cases well-defined, severe pathological conditions (head trauma, intracranial tumour, metabolic emergency, etc.) stand in the background of the development of cortical blindness (CB). The cause of such cortical loss of vision which develops without preceding or accompanying acute disease, however, sometimes remains unkwown. Nevertheless, in cases when CB develops in a patient suffering from some, otherwise not severe chronic disease which requires continuous treatment, the possibility always arises that either the chronic disease itself, or the chronic treatment may have contributed to the pathogenesis of visual impairment. In the case presented here CB developed in an infant who suffered from congenital adrenal hyperplasia (CAH) and, during an acute episode of infection, received steroids in unusually high doses. Therefore, the possibility that in this case the CB developed was related either to the CAH or to the treatments administered may probably be worthy of attention.

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## CASE REPORT

A 10-month-old boy was admitted to a county hospital because of fever, watery diarrhoea and repeated vomiting. He was known to suffer from the salt-losing form of 21-hydroxylase deficiency diagnosed in the neonatal period on the basis of 17-OH progesterone profile. Before admission he received daily 18.75 mg cortisone acetate (47 mg/$m^2$ body surface area) and 0.1 mg fludrocortisone (0.25 mg/$m^2$ body surface area). Repeated determinations of serum 17-OH progesterone profile indicated suitable suppression; serum potassium levels were in the normal range.

On admission metabolic acidosis (pH: 7.2;BE: -14 mEq/l) and mild hyponatraemia (serum Na: 133 mEq/l) were detected. Bacterial cultures were negative and an enteritis of viral origin was suspected. The doses of steroid therapy were substantially increased, i.e. for 3 days the infant was given 75 mg prednisolone sodium succinate and 0.15 mg fludrocortisone daily, then the previous maintenance therapy was reintroduced. Intravenous fluid therapy could be stopped by the end of the 1st week and the infant was discharged on the 12th day of hospitalization. At home the parents soon noticed that the infant could not recognize them or the familiar objects of his environment. Next day the infant was admitted to our department.

On admission an apparently healthy infant was seen who did not react to any visual stimulus, whilst the pupillary reflexes were intact. Fundoscopy was normal. No other neurological signs were seen and the CSF was normal. Visual evoked response investigation demonstrated a severe lesion of the visual system with preservation of some electrical activity. Brain scintigraphy showed normal activity, but the CT scan revealed severe bilateral occipital hypodensity. The diagnosis of cortical blindness was established.

On admission blood pressures were markedly elevated (180/110 mm Hg). Blood gases, electrolytes (serum Na: 141 mmol/l; serum K: 4.6 mmol/l), vaneline mandelic acid excretion and plasma aldosterone values were normal. The hypertension proved to be transient and after three weeks of combined antihypertensive therapy given in decreasing doses no further medication was needed. The congenital adrenal hyperplasia was treated by daily 18.75 mg cortisone acetate. While the infant was hypertensive no mineralocorticoid therapy was given, serum sodium level was maintained in the normal range by oral sodium supplementation alone. In the normotensive state the original steroid therapy was reintroduced.

By the end of the one-year follow-up period remarkable improvement of vision was seen, the little boy became able to differentiate the objects in his immediate proximity. He was normotensive and the CT showed narrowing of the region of hypodensity. No neurological signs were seen; visual evoked potentials showed improvement (higher amplitudes and shorter latency of the response), but some evidence of the lesion could still be demonstrated.

## DISCUSSION

Even in the paediatric age group a variety of pathological conditions may result in sudden bilateral loss of vision /2/. Also CB is a well-known entity /5/ and had also been reported in association with various metabolic emergencies /6/.

In the case presented here CB developed following an enteritis of medium severity and was accompanied, or at least followed, by severely elevated blood pressure. Most unfortunately, at first hospitalization, when occipital atrophy had presumably taken place, blood pressures were not recorded. It is possible that before the institution of emergency therapy the child might have had an addisonian like situation with low blood pressure for some time and/or severe hyponatraemia giving rise to cerebral oedema. Certainly, the brain damage might have had this background; however, it seems to be more probable that the simultaneous occurrence of elevated blood pressure measured later on and CB is not a mere coincidence. Thus, the CB of the patient seems to be related to hypertensive encephalopathy (the lack of papilledema may be explained by the fact the fontanelle was still open).

Intracranial hypertension is a well-known complication of steroid therapy /1/ and CB has also been described in children suffering from CAH and treated with steroids /3,4/. In the case presented the duration of maintenance steroid therapy prior to the hospitalization was short and the drugs were given in conventional doses. For the incriminate three days of the first hospitalization, however, corticoids were given in unusually high doses. If the dose relation for equal effects between prednisolone and cortisone, and between prednisolone and cortisol are assumed to be 1:5 and 1:4, respectively, the steroid doses given would correspond to approximately 300 mg of cortisol or 375 mg of cortisone. This means about 20 times the dose administered before the 1st hospitalization. The elevated steroid dosage might have contributed to the pathogenesis of hypertension and CB.

It is well-known that in patients requiring steroid therapy the doses administered should be increased in stress situation.

The severe complication observed in the present case, however, suggests that high doses of steroids, and especially mineralocorticoids, may have serious consequences.

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