

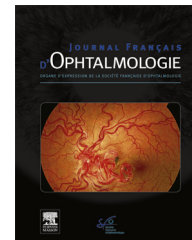


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ORIGINAL ARTICLE

Evaluation of the visual function of patients with locked-in syndrome: Report of 13 cases



Évaluation de la fonction visuelle chez des patients locked-in syndrome : à propos de 13 cas

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KEYWORDS

Locked-in syndrome;
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VIth nerve palsy;
Keratitis

Summary Although visual function is thought to be preserved in patients with locked-in syndrome (LIS), enabling them to communicate through vertical or lateral eye movements or blinking of the upper eyelid, nothing is known about the actual visual function of patients with LIS. The goal of this study is to evaluate the visual function of patients with LIS which may enable better evaluation of the state of consciousness of these patients. Patients with LIS seen in a single neurovascular unit of a tertiary center between 1997 and 2013 were retrospectively reviewed. Each patient had a specialized neuro-ophthalmological evaluation under optimal environmental conditions (light, contrast, examination distance, head position, best ergonomic adaptation, and establishment of a means of communication with help from the patient's friends/family). Visual acuity, extraocular eye movements, confrontational visual field, slit lamp and fundus examination were performed. Thirteen patients (6 M/7F) were included in this study. Mean visual acuity was 20/60. Oculomotor examination was abnormal in 77%. Forty-six percent of patients presented binocular diplopia mainly related to a VIth nerve palsy. One patient presented complete ophthalmoplegia. Forty-six percent of patients had nystagmus responsible for oscillopsia (oculopalatal tremor). An abnormal visual field was observed in 17% of patients, and abnormal pupillary light response leading to photophobia was present in 22% of patients. Keratitis or dry eye syndrome was present in most patients and was a major cause of pain and visual impairment. Our results suggest that the visual function is impaired in all patients with

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LIS. This impairment is multifactorial including mostly binocular diplopia or oscillopsia but also refractive errors, dry eye syndrome, keratitis or visual field defect. This altered visual function may alter the ability of the patient to interact with his environment and lead to underestimation of their state of consciousness. An ophthalmologic evaluation would allow for improvement of these patients' comfort, their ability to communicate, and the assessment of their state of consciousness.

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MOTS CLÉS

Locked-in syndrome ;
Diplopie ;
Oscillopsie ;
Nystagmus ;
Paralysie du VI^e nerf
crânien ;
Kératite

Résumé Il est communément admis qu'il n'y a pas d'atteinte des voies visuelles chez les patients *locked-in syndrome* (LIS). Cependant aucune étude ne rapporte d'évaluation de la fonction visuelle chez ces patients. L'objectif de cette étude était d'évaluer la fonction visuelle chez des patients LIS et à travers celle-ci, leurs capacités de communication et leur degré de conscience. Étude monocentrique, rétrospective incluant tous les patients atteints de LIS, vus dans une unité neurovasculaire entre 1997 et 2013. Chaque patient bénéficiait d'un examen neuro-ophtalmologique après contrôle optimal des conditions d'examen (luminosité, contrastes, positionnement, établissement d'un code de communication avec l'aide de l'entourage). L'acuité visuelle, l'oculomotricité, un champ visuel par confrontation, un examen à la lampe à fente avec fond d'œil étaient relevés. Treize patients ont été inclus dans l'étude. L'acuité visuelle moyenne était de 20/60 (échelle de Snellen). Il existait 77 % d'atteintes oculomotrices, 46 % de diplopie par paralysie du VI. Un cas d'ophtalmoplégie, 46 % de nystagmus responsables d'oscillopsies. Dix-sept pour cent d'atteintes du champ visuel et une anomalie de l'examen pupillaire dans 22 % des cas. Les kératites par syndrome sec étaient présentes chez la plupart des patients et constituaient leur principale plainte fonctionnelle ophtalmologique. Ces résultats suggèrent que la vision est altérée pour la majorité des patients souffrant de LIS. Les causes de baisse de vision sont multiples : diplopies, oscillopsies, kératites par syndromes secs, atteintes du champ visuel. Une évaluation ophtalmologique permettrait chez ces patients afin d'améliorer leur confort, leurs capacités de communication et l'évaluation de leur état de conscience.

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Introduction

Locked-in syndrome (LIS) is a rare condition, first described in 1966 by Plumm and Posner [1]. It associates preservation of eye opening, basic cognitive abilities, severe hypophonia or aphonia, quadriparesis or quadriplegia. The main mode of communication is limited to vertical or lateral eye movements or blinking of the upper eyelid [2,3]. The only way to communicate or establish interactions with the environment relies on eye-coded communication strategies. Visual function is thus a major issue. Abnormal eye movements with preservation of voluntary vertical eye movements and upper eyelid closure, and variable loss of voluntary and reflex horizontal eye movements (ocular bobbing, opsoclonus myoclonus, abolition of vestibulo-ocular, oculocephalic reflex) have been reported in LIS. However, the actual visual function of these patients, including visual acuity, visual field, ocular surface integrity and extraocular eye movements, has never been evaluated. The aim of this study is to describe ophthalmological manifestations of LIS, and to evaluate the global visual function of these

patients, which is often the only way to assess the state of consciousness of these patients.

Material and methods

Medical records of patients with LIS seen in a single tertiary center between 1997 and 2013 were retrospectively reviewed. The same examiners including a neurologist, an ophthalmologist and an orthoptist evaluated every patient. The first step was to identify the communication code that had been established between the patient, and the family or the nursing staff. Clinical ophthalmological evaluation was performed, including visual acuity, extraocular eye movements, pupillary light reflex, slit lamp examination (portable slit lamp device), fundus examination, and confrontational visual field. Eyelid function including eye closure and levator function were also assessed. Refraction measure was performed, using a portable refractometer whenever possible. Several variations of the environmental conditions, including light, contrasts, and distance of examination, head

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