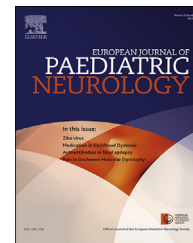




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Original article

Comparison of the detectability of UBOs in Neurofibromatosis Type I patients with proton density-weighted and FLAIR sequences in 3T MRI

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ABSTRACT

Objective: In NF 1 patients, significant numbers of so-called unidentified bright objects (UBOs) can be found. The aim of the study was to investigate whether the detectability of UBOs increases at 3T by comparing Proton density-weighted images (PDw) with fluid-attenuated inversion recovery (FLAIR) sequences.

Patients and methods: A total of 14 NF1 patients (7 male, 7 female, between 8 and 26 years old, mean age 15.4 years) were examined by a 3T magnetic resonance scanner. The presence of UBOs was evaluated on PD-w and FLAIR images by 4 evaluators. Detectability was rated by a three-point scoring system: lesions which were “well defined/detectable”, “suspicious” or “detected after a second look”. The Wilcoxon signed-rank test was used for comparisons between the raters. The level of significance was $P < 0.05$.

Results: Significantly more lesions were marked as “well defined/detectable” in the PD-w Sequence compared to FLAIR at 3T ($P < 0.001$ for all four evaluators together, as well as for each evaluator separately). In particular, PD-w proved to be superior for detecting UBOs located in the medulla oblongata, dentate nucleus and hippocampal region, regardless of the level of the raters' experience.

Conclusion: This is the first study which compares FLAIR and PD-w at 3T for the diagnosis of UBOs in NF1. At this field strength significantly more UBOs were detected in the PD-w compared to FLAIR sequences, especially for the infratentorial regions. As UBOs occur at very early stages of the disease in patients with suspected NF1, PD-w might aid in the early diagnosis when using 3T scanners.

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1. Introduction

Neurofibromatosis Type 1 (NF1) is an autosomal-dominant congenital disease, which first manifest in the early stages of childhood. With a prevalence of 1 in 2000 to 1 in 3300, NF1 is one of the most common hereditary mutations and the most common neurocutaneous syndrome. Diagnostic features of NF1 have been formulated by the National Institutes of Health Consensus Development Conference (NIHCDC criteria).¹ The clinical presentation in NF1 is variable, comprising neurological or psychomotoric disorders with intellectual impairment as well as osseous lesions like dysplasia, excavations of the vertebral bodies or macrocephaly. Furthermore, NF1 patients often suffer from

neoplasia such as optic nerve gliomas. Neurocutaneous lesions occur frequently in these patients, including café au lait spots, axillary or groin freckling, Lisch nodules and the eponymous neurofibromas. 2 or more NIHCDC criteria are required for the diagnosis of NF1.

Magnetic resonance imaging (MRI) is the investigation of choice to rule out involvement of the brain tissue. In NF1 patients, significant numbers of so-called unidentified bright objects (UBOs) can be found in brain imaging, with predilection sites at the basal ganglia and the dentate nucleus (see Fig. 1). They occur as non-enhancing lesions of increased intensity on T2 weighted images without any mass effect. The nature of the abnormalities is uncertain. In the past it has been assumed that they might be hamartomas or very slow growing gliomas.² The most recent hypothesis is that they

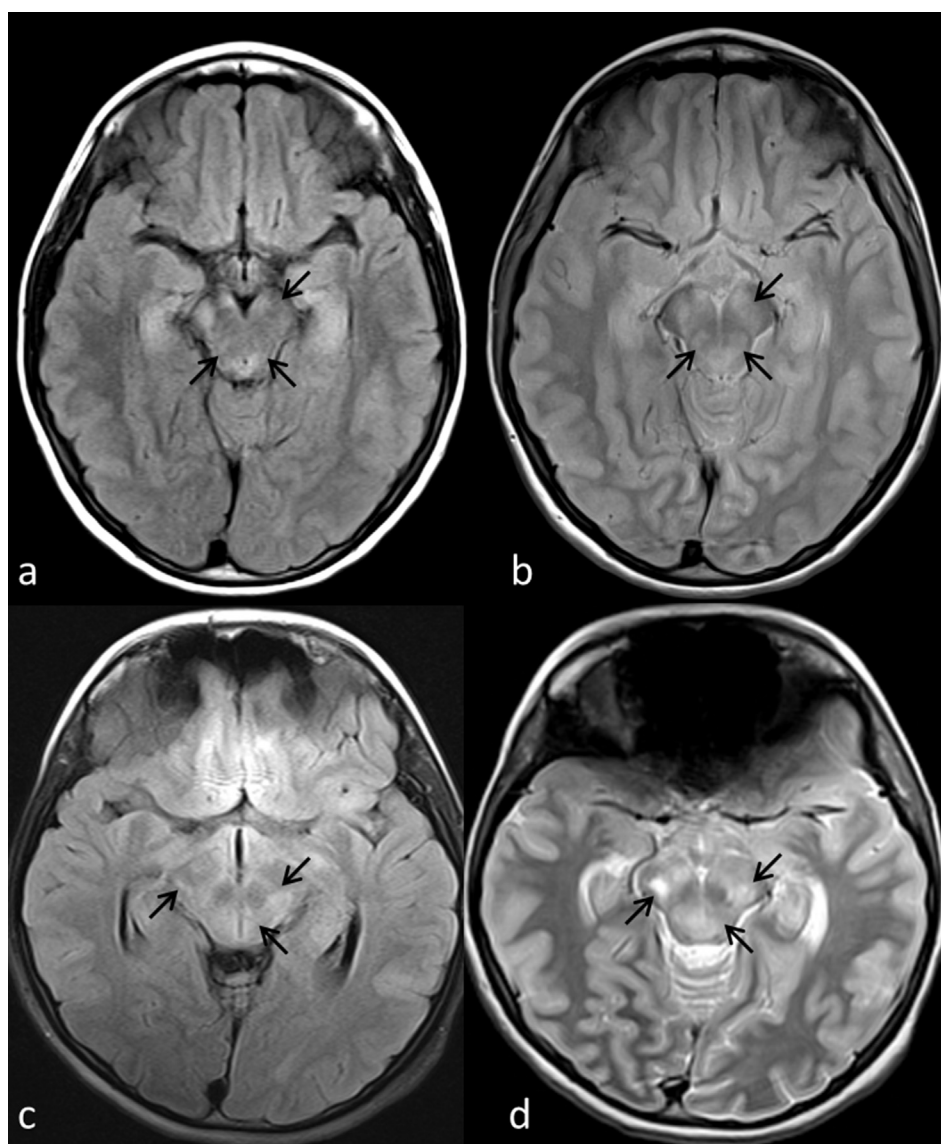


Fig. 1 – FLAIR (a + c) and PDw (b + d) images in axial orientation of two patients with NF 1: In the first patient (upper row a+b) hyperintense lesions (arrows) can be well detected in the PDw images (b) located in the dorsal parts of the midbrain as well as in the crus cerebri of the left side compared to the FLAIR images (a). In the second patient (lower row) UBOs (arrows) at the mesencephalic region are well seen in the PD images (d) and can only be suggested at the FLAIR (c) images at a second look.

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