Response to: 'Standard dose of ustekinumab for childhood-onset deficiency of interleukin-36 receptor antagonist' by Cherqaoui *et al*

The case reported by Cherqaoui *et al*¹ on ustekinumab in the treatment of deficiency of interleukin-36 receptor antagonist (DITRA) is interesting in several aspects.

The authors describe intrathecal elevation of proinflammatory cytokines, which is a novel and intriguing finding. The mutation in their case is identical to that of the Dutch/Moroccan patient we described. The total absence of any clinical disease activity in a genetically affected younger sister of the patient further supports the variable expression of identical *IL36RN* mutations as reported by Cherqaoui *et al*.

As observed in our cases, the patient was treated with several drugs with an absent or only transient improvement.² At variance with our previous experience, the use of ustekinumab at standard dose in monotherapy was able to completely control the clinical picture, without the need for any topical steroids. These observations underline the clinical spectrum of disease severity associated with DITRA. In our previous experience, both patients displayed an initial response at standard doses but displayed a tendency to mild/moderate relapses in proximity to the scheduled doses. According to the few cases so far described, it is therefore conceivable that ustekinumab should be considered among the first-line treatments for this severe condition. As suggested by Cherqaoui *et al*, a standard dose of 0.75 mg/kg every 12 weeks could be sufficient to completely control disease flares and should therefore be used as an initial approach. However, higher doses at more frequent intervals could be necessary to achieve a complete remission in patients with a partial clinical response, with a good safety profile.

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