Clinical case descriptions of the two patients with Grade 5 infectious AE, and two patients with Grade 4 infectious AE.

The first was a nine year old female with NMDAR encephalitis requiring intensive care, corticosteroids and IVIG. Due to therapy failure, the patient received rituximab 28 days after first line therapy. The patient developed CMV colitis a month after rituximab whilst still on oral prednisolone. Endoscopy was complicated by bowel perforation and subsequent death (AE Grade 5, directly attributable to immune suppression). The second patient had severe NMDAR encephalitis requiring intensive care. The patient received plasma exchange on days 10-15 followed by IVIG supplementation, intravenous methylprednisolone on day 25, then rituximab was started on day 48. The patient made significant neurological improvements but developed fatal Staphylococcal toxic shock syndrome 24 days after the second dose of rituximab (AE Grade 5, possibly related to immune suppression). The third patient was a ten year old female with neuropsychiatric lupus who did not respond to intensive care, corticosteroids and IVIG, and therefore received rituximab and concomitant cyclophosphamide. The patient developed CMV retinitis and has been left with bilateral visual disability (visual acuity 20/70) (AE Grade 4, directly related to immune suppression). The fourth patient was a six year old male with NMDAR encephalitis treated with corticosteroids, IVIG, and concomitant rituximab who developed severe shock and secondary hypoxic brain lesions three days after immune therapy. Although the precise aetiology for his decompensation was not clear, the possibility of a delayed allergic reaction or a septic event due to an unidentified infection was considered. The patient was left quadriparetic secondary to the diffuse brain injury (AE Grade 4, possibly related to immune suppression).