

Author, Year [reference] Source	Sex (years) Genotype (AAT serum in mg/dL)	Clinical data	Effect of augmentation therapy
Smith et al, 1987 [62] Rochester, US	Case 1. F (65). ZZ (40)	Extensive ulcerative relapsing panniculitis requiring hospitalization	After the failure of Indomethacin, Prednisone, Azathioprine, Ketoconazole and Dapsone, a single infusion of AAT (60 mg/Kg/week) was applied in 2 severe recurrences, with significant clinical efficacy.
Smith et al, 1987 [62] Rochester, US	Case 2. F (36). ZZ (20)	Extensive ulcerative relapsing panniculitis requiring hospitalization	After the failure of Prednisone and Dapsone, 7 weekly infusions of AAT were successfully applied together with Dapsone and Prednisone.
Furey et al. 1996 [63] Chicago, US.	Case 3. F (62). ZZ. (24)	Extensive ulcerative relapsing panniculitis with massive edema (in legs, thighs and abdomen) requiring hospitalization	A single infusion of AAT (60 mg/ Kg) was provided. The response was dramatic. Within 24 hours, the patient could walk; and after 3 weekly infusions, both massive edema and panniculitis were resolved.
O'Riordan et al, 1997 [64] Chicago, US	Case 4. F (62). ZZ (23)	Severe ulcerative relapsing panniculitis	Panniculitis was completely resolved with 3 weekly doses of IV AAT (60/mg/Kg). Skin lesions recurred 6 months later but were resolved with 2 additional infusions, once levels of AAT were over 50 mg/dL.
Chowdhury et al 2002 [65] Cardiff, UK.	Case 5. F (33). ZZ (46)	Rapidly progressive panniculitis, septic shock and multiple life-threatening complications	Augmentation therapy was life-saving. Prolastin® quickly reduced inflammation within 24 hours. Since then, the patient has been feeling well, inflammation-free, under a treatment of 6 g/ week of Prolastin®.
Kjus T et al, 2003 [66]. Oslo, Norway	Case 6. F (21). ZZ (30)	Ulcerated chronic relapsing panniculitis.	After the failure of dapsone and doxycycline, the patient was successfully treated with i.v. infusions of Prolastin® (60/mg/Kg) once a week. “All lessons resolved within 2 days; reappeared after 4 weeks, and again disappeared 1-2 days after Prolastin®. At present, well controlled with infusions every 4th week”
Gross B [67]. 2009. Frankfurt, Germany.	Case 7. F (31). ZZ (22)	Ulcerated relapsing panniculitis. <i>Z-type AAT polymers were found in skin, supporting the hypothesis of the pro-inflammatory role of polymers.</i>	The long-term use of intravenous AAT augmentation therapy (Prolastin®) during 3 years reduced both the frequency and severity of panniculitis.