Table 1. Neurologic manifestation of 42 patients with refractory neurobehçet's disease treated with biologic therapy.

Parenchymal subtype, n (%)	34 (81.0)
-Hemiparesis	8 (19.1)
-Polineuropathy	8 (19.1)
-Encephalopathy	6 (14.3)
-Cognitive impairments	4 (9.5)
-Optic neuropathy	4 (9.5)
-Ophthalmoparesis	4 (9.5)
-Other cranial nerve involvement	3 (7.1)
-Hemihypoesthesia	3 (7.1)
-Cerebellar dysphasia	1 (2.4)
-Cerebellar involvement	1 (2.4)
-Non-steroidal psychosis	1 (2.4)
Non-parenchymal subtype, n (%)	17 (40.5)
-Aseptic meningitis	12 (28.6)
-Thrombosis	4 (9.5)
-Intracranial hypertension	1 (2.4)

Figure 1. Neurological clinical response to biological therapy.

Primary failure was observed in 16 (38.1%) patients due to inefficacy (n=11, 68.8%) or adverse effects (n=5, 31.3%). Similarly, causes of secondary failure (n=6, 14.3%) were inefficacy (n=5, 83.3%) and adverse effects (n=1, 16.7%). No serious adverse effects were observed.

Conclusion: BT, especially monoclonal anti-TNF drugs, seems to be effective and safe in refractory NBD.

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POS1372

EVALUATION OF E148Q AND CONCOMITANT AA AMYLOIDOSIS SECONDARY TO FAMILIAL MEDITERRANEAN FEVER AFTER ADJUSTED CLINICAL-DEMOGRAPHIC CHARACTERISTICS

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Pediatrics, London, Canada; ⁴Gentera Biotechnology, Biotechnology, Istanbul, Turkey; ⁵Epigenetic Health Solutions, Unit of Nephrology, Ankara, Turkey

Background: Amyloid A (AA) amyloidosis, previously known as secondary or reactive amyloidosis, is a long-recognized severe complication of some chronic inflammatory diseases. The pathogenesis and risk factors for amyloidosis in Familial Mediterranean Fever (FMF) remain partially understood (1). The development of AA amyloidosis is dependent on ethnicity and country of residence (2). In the pre-colchicine era, renal AA-amyloidosis was largely reported patients of Turkish (67%) and Sephardic Jewish ancestry (26.5%) (2,3). Currently it's well known that the MEFV M694V variant associated with high risk of amyloidosis however, mutations on exon 2, specifically E148Q variant remained controversial.

Objectives: To evaluate the E148Q mutation variant and concomitant AA Amyloidosis secondary to FMF after adjusted clinical-demographic characteristics.

Methods: Patients were recruited from the renal unit at Epigenetic Health Center outpatient clinic in Turkey between September 2003 and February 2020. Patients who had biopsy confirmed FMF related AA amyloidosis were included the study. Tel-Hashomer criteria were applied in the diagnosis of FMF. The clinical characteristics of FMF patients and medication history were recorded by the physician at the time of registry entry. All patients had detailed baseline assessment of clinical features, renal functions, genetic testing, histopathological diagnosis of amyloidosis, and treatment received. We performed multiple comparisons according to the age of diagnosis, demographic features, disease phenotype, allele frequency, type of mutation and mortality. Statistical analysis was performed with Statistical Package of Social Science (SPSS) for Windows, version 15.0 (SPSS Inc, Chicago, IL).

Results: Our registry consists of 195 patients with a diagnosis of AA amyloidosis. Complete information on 169 patients (lost to follow up, n=26) were included. The median age was 36 (19-49) years; male/female ratio was 1.6 (104/65). The median follow-up duration was 15.0 years (4-17 years). There were 101 patients diagnosed with FMF <18 years of age and 68 patients diagnosed ≥18 years of age. All participants developed renal amyloidosis before the age of 32 years. Family history of FMF was documented in 56 patients (33.1%) and family history of amyloidosis was present in 41 patients (24.3%). The three most common clinical symptoms were fever (84.6%), abdominal pain (71.6%) and arthritis (66.9%). During the follow-up, 5 patients started dialysis treatment and 9 patients had kidney transplantation. The most common allele frequency across patients was M694V (60.6%), E148Q (21.4%) and M680I (10.3%). The most frequent mutations were M694V/M694V (63.3%), M694V/E148Q (20.8%) and E148Q/E148Q (15.8%). During the follow up period, 15 patients (10 male, 5 female) died. In those that died, the mutations in 14 had M694V/M694V and one demonstrated E148Q/E148Q.

Conclusion: Patients with FMF related AA amyloidosis have an increased risk for mortality. This study confirmed the association between M694V and FMF-associated AA amyloidosis, which has been reported in many studies. Close clinical follow-up and further evaluation of patients with the E148Q mutation is warranted specifically if residing in FMF endemic areas. The possible relationship between E148Q and AA amyloidosis need to be confirmed in other cohorts.

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POS1373

DEFINING EAR CHONDROITIS: DATA FROM 685 PATIENTS WITH RELAPSING POLYCHONDROITIS

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Background: Ear chondritis is often considered the pathognomonic feature of relapsing polychondritis (RP). Although painful redness and swelling of the pinna and a resultant cauliflower ear are universally recognized as chondritis, the complete spectrum of symptoms associated with ear chondritis have not been well described.