Behçet’s Disease: Drug pathway

Less complex disease

Mucocutaneous disease (90% of patients)

+/- arthropathy arthritis

Behçet’s Eye disease (70% of patients)
Acute Flare (hypopyon panuveitis with occlusive retinal vasculitis)

Other major organ disease (e.g., Central nervous system disease, peripheral neurological disease, major vessel thrombosis (20% of patients)

Syndrome

Severe disease

Drugs

Initial therapy

Oral colchicine 500 microg bd
Topical steroid (mouthwash, gel, spray)
Topical non-steroidal (anti-inflammatory mouthwash)
Triple mouthwash
Antibiotics

Initial acute therapy

Steroid
Topical, Intraocular; Intravenous (3x1000mg); Oral (prednisolone 1mg/kg/day)
Maintenance therapy
oral steroid
azathioprine 2mg/kg/day
cyclosporin 2.5-5mg/kg/day

TNF inhibitor

Initial therapy

Infliximab 5mg/kg for 4 doses then swap to either adalimumab 40mg eow or etanercept 50mg once a wk
Stop therapy after 24 months. Restart if flare (70% flare within 3 months)

Step up therapy

Azathioprine 2.5 mg/kg (up to dose 200mg/day) or Tacrolimus up to 4mg bd (dosed on plasma trough concentration)
or Steroid – variable daily, monthly, yearly intramuscular (120mg depomedrone up to 6 times/yr) Intravenous (1000mg depomedrone once a yr) oral variable dose

Step up therapy

Cyclophosphamide (intravenous pulses 10mg/kg at increasing intervals 0, 2, 4, 8, 12, wks)

Biological agent

Initial therapy

Infliximab 5mg/kg for 4 doses then swap to either adalimumab 40mg eow or etanercept 50mg once a wk. Stop therapy after 6 mnths. Restart if flare (70% flare within 3 mnths)

The bottom line: mucocutaneous disease -
75% patients taking baseline drugs
20% patients on step up therapy
5% patients requiring TNFi

The bottom line: eye disease -
85% patients taking baseline drugs
10% patients requiring TNFi
5% patients requiring IFN/rituximab

The bottom line: other major organ flare -
60% patients taking steroid/azathioprine
30% patients on cyclophosphamide
10% patients requiring a biologic agent

The bottom line: syndrome -
80% patients taking baseline drugs
20% patients taking additional therapy