

Short Term Prophylaxis

Minor Manipulations

If plasma-derived C1 inhibitor (pdC1INH) immediately available:

- No prophylaxis needed

If pdC1INH not available:

Prophylaxis for five days before and two to 5 days post event

- Danazol (avoid during first two trimesters of pregnancy; 2.5-10 mg/kg/day, maximum 600 mg daily)
- Stanozolol 4-6 mg/day

Major Procedures or Intubation:

Plasma-derived C1 inhibitor (pdC1INH)

Give one to six hours before procedure*
(optimum dose not yet established – see text)
Second dose of pdC1INH should be immediately available

If pdC1INH not available:

Danazol prophylaxis as per minor and Solvent/detergent treated plasma (SDP; if not available, then fresh frozen/frozen plasma but less safe than SDP) one to six hours before procedure*
10 ml/kg; 2-4 units (400-800 ml) for an adult
***as close to procedure as feasible**

Long Term Prophylaxis

Plasma-derived C1 Inhibitor

If failing on demand therapy, then continuous pdC1INH prophylaxis twice weekly

Androgens

Danazol (≤ 200 mg/day)
Or Stanozolol (≤ 2 mg/day)
(use lowest effective dose)

Antifibrinolytic Agents

Less effective than androgens
Tranexamic Acid (TA)
20-50 mg/kg/day split bid or tid
(3-6 g/day maximum)
(Epsilon aminocaproic acid, EACA if TA not available)