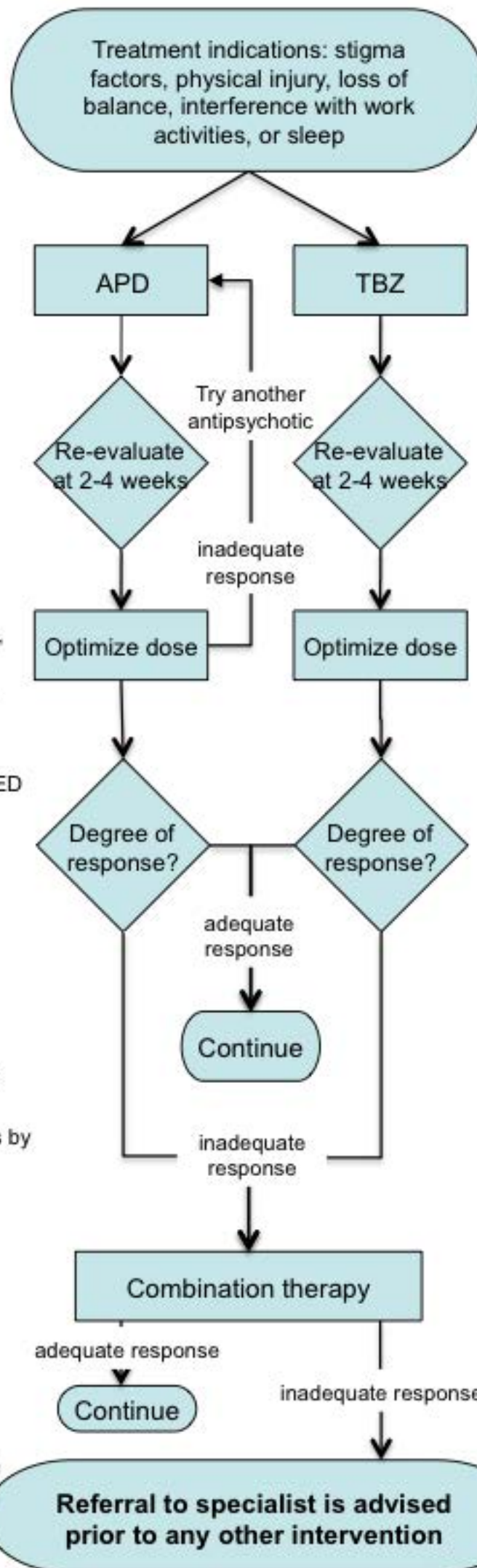


Algorithm for the treatment of chorea in Huntington's disease



Tetrabenazine (TBZ)

Avoid if co-morbid:

- psychosis
- active depression
- aggressive behaviors
- non-compliance

Step 1. Start with 12.5 mg/day

Elimination half life varies greatly among individuals from 2-8 hours, and will ultimately require 2-4 doses per day. Because chorea abates with sleep, bedtime dosing is usually not helpful unless chorea interferes with sleep.

If used with SSRI avoid fluoxetine, paroxetine which can prolong half-life (see text)

Goal is to decrease chorea severity, not to eliminate it.

Step 2. Dose optimization

Go slow, by 12.5mg/day increments. Though the manufacturer suggests 1-week dosing intervals, most experts use 2 or more weeks before increasing dose. Reassess for response and side effects at each dose increment.

Therapeutic dosage variable (12.5-75 mg/day). Referral to specialist advised for higher dosing.

Side effects are dose related, with higher incidence of side effects when used with SSRI, AED, or APD:

- sedation
- depression
- suicidal behaviors
- Parkinsonism
- apathy
- swallowing impairment
- akathisia
- neuroleptic syndrome

Stop the drug for suicidal behavior.

Decrease dosage for control of other side effects.

Add or increase SSRI for TBZ-associated depression.

Step 3. Combination therapy

Add BZD if anxiety-related

Add APD with attention to increased side effects

Antipsychotic (APD)

First choice if co-morbid:

- psychosis
- active depression
- aggressive behaviors
- non-compliance

Step 1. Start with low dose

olanzapine	(2.5-10 mg)
risperidone	(0.5-2 mg)
haloperidol	(0.5-2 mg)
quetiapine	(25-200 mg)
tiapride*	(50-200 mg)
aripiprazole	(2-15 mg)
sulpiride*	(100-600 mg)

*tiapride, sulpiride available in Europe

Advise twice daily dosing to minimize side effects and at least a 2-week interval prior dose increase

Step 2. Dose optimization

Go slow and gentle: goal is to decrease, not eliminate, chorea

Reassess response and side effects at each dose increment

Side effects are dose related; higher incidence when added to TBZ, SSRI, AED

Side effects:

- sedation
- Parkinsonism
- apathy
- cognitive impairment
- akathisia (motor and psychic restlessness)
- metabolic syndrome
- swallowing impairment
- tardive dyskinesia
- neuroleptic syndrome (can occur with rapid dose escalation or lowering)

Frequency of specific side effects varies by drug (see text).

Step 3. Combination therapy

Add BZD if anxiety-related

Add TBZ with attention to increased side effects

Abbreviations

AED	mood stabilizing antiepileptic drug
APD	antipsychotic
BZD	benzodiazepine
SSRI	serotonin reuptake inhibitor
TBZ	tetrabenazine