HD Chorea Algorithm: Patient and Family Summary

Experts believe that drug treatment is appropriate when chorea causes embarrassment, interferes with employment, balance, sleep, or if it causes injury to you or your care partner. If chorea is a part of your HD, you and your care partner should talk about it before the visit. Tell you story, and if after discussion with your doctor, a drug trial is appropriate, it is important for both of you to remember that the goal is to decrease chorea, not to eliminate it, and that drug response is different from patient to patient.

Step 1: Some experts prefer to start with an antipsychotic drug (like risperidone (Risperdal) or olanzapine (Zyprexa), and others prefer Xenazine (tetrabenazine). The best drug for you will depend on other HD related problems you may have. For instance, if you are depressed, or have aggressive behaviors, or psychotic symptoms (like paranoid delusions) the best choice is an antipsychotic drug. In other situations many experts prefer Xenazine. As a relatively new drug, your local doctor may not know how to prescribe it, or what programs are offered to those who can't afford it. It may be best to bring information found on the web at the Xenazine Information Center.

Step 2: Finding the best dosage of either type of drug will take multiple visits. Don't skip any! Most experts use at least a 2-week time interval. Both you and your care-partner should watch for benefit: check to see if chorea is better the hours after taking a dose, then gets worse before the next dose, or whether you have been able to resume activities which hadn't been able to do before treatment, which can help the doctor manage dosage. Equally important, watch for side effects and contact your doctor for:

- Depression or suicidal thoughts
- Daytime sleepiness or insomnia
- More trouble with concentration
- Restlessness (physical or mental)
- Loss of interest in usual activities

Many side effects can be controlled by decreasing the dose of drug except for suicidal thoughts when using Xenazine, it usually isn't necessary to stop the drug.

Step 3: If one drug doesn't give satisfactory results, some experts will use an antipsychotic and Xenazine together. Or if anxiety is making the chorea worse; experts frequently add a drug like clonazapam (Klonopin) or lorazepam (Ativan). More rarely they might add amantadine, a drug more often used for Parkinson's disease.

Step 4: Best dosing of drugs often changes over time and stage of disease. Your doctor will decide when whether a trial of decreasing drug dosage is advised.

Summary by LaVonne Goodman M.D. based on Groves M, van Duijn E, Anderson K, Craufurd D, Edmondson MC, Goodman N, van Kammen DP, Goodman L. An International Survey-based Algorithm for the Pharmacologic Treatment of Irritability in Huntington's Disease. *PLoS Curr* 2011;3():RRN1259.

Algorithm for the treatment of chorea in Huntington's disease

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).

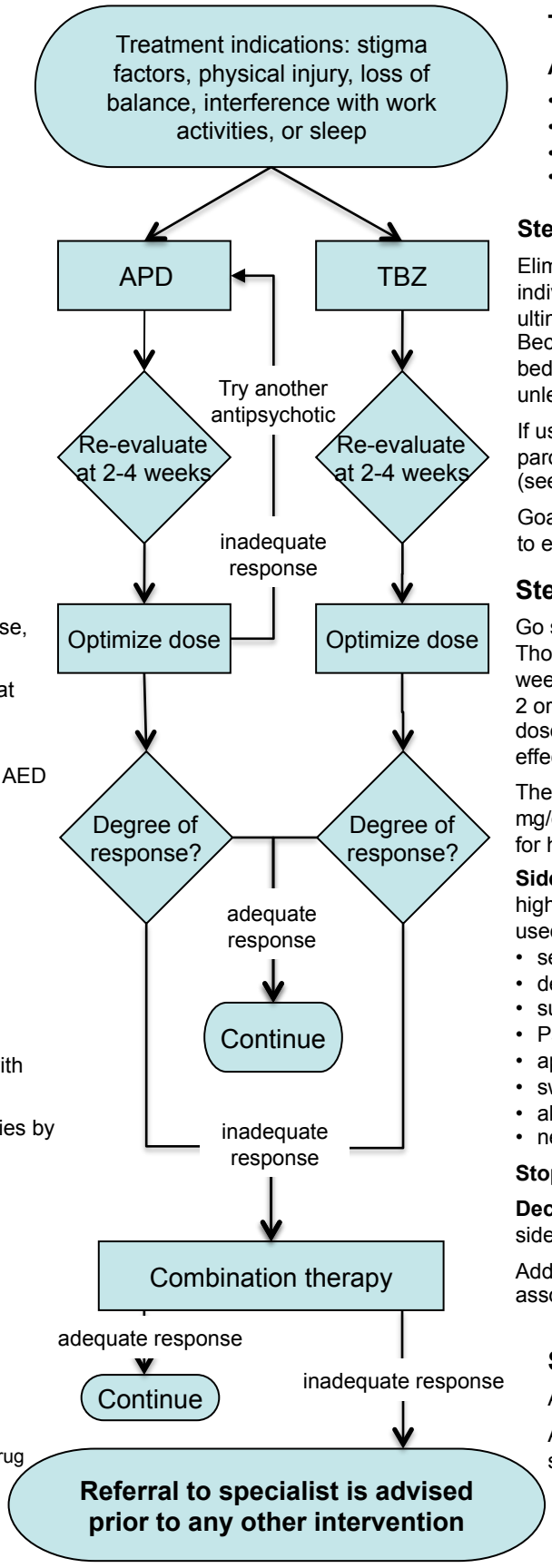
Step 3. Combination therapy

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



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

Groves M, van Duijn E, Anderson K, Craufurd D, Edmondson MC, Goodman N, van Kammen DP, Goodman L. An International Survey-based Algorithm for the Pharmacologic Treatment of Irritability in Huntington's Disease. PLoS Curr 2011;3():RRN1259.

Anderson K, Craufurd D, Edmondson MC, Goodman N, Groves M, van Duijn E, van Kammen DP, Goodman L. An International Survey-based Algorithm for the Pharmacologic Treatment of Obsessive-Compulsive Behaviors in Huntington's Disease. PLoS Curr 2011;3():RRN1261.

Burgunder JM, Guttman M, Perlman S, Goodman N, van Kammen DP, Goodman L. An International Survey-based Algorithm for the Pharmacologic Treatment of Chorea in Huntington's Disease. PLoS Curr 2011;3():RRN1260.

Available for print at:
WeHaveAFace.org