

Supplementary material to: Aggarwal A, Bhatt M. The pragmatic treatment of Wilson disease. Movement Disorders Clinical Practice, 2014.

1. Global Assessment Scale for Wilson Disease (GAS for WD) Score Sheet
2. GAS for WD rating instructions

Global Assessment Scale for Wilson Disease: Score Sheet (Aggarwal A, Bhatt M. MDCP, 2014)

Patient Identifier Number:

Rater Initials								
Date (DD/MM/YY)								
TIER 1: DOMAINS								
1. Liver (0-5)	L	L	L	L	L	L	L	L
2. Cognition & behavior (0-5)	C	C	C	C	C	C	C	C
3. Motor (0-5)	M	M	M	M	M	M	M	M
4. Osseomuscular (0-5)	O	O	O	O	O	O	O	O
TIER 2: ITEMS								
1. Wilson's facies (0-4)								
2. Scholastic Performance (0-4)								
3. Depression (0-4)								
4. Psychosis (0-4)								
5. Dystonia (0-4)								
6. Tremor (0-4)								
7. Chorea (0-4)								
8. Parkinsonism (0-4)								
9. Speech (0-4)								
10. Swallowing (0-4)								
11. Salivation (0-4)								
12. Posture and Gait (0-4)								
13. Kayser-Fleischer rings (0-4)								
14. Uncommon (1 for each; max 4)								
Tier 2: Total Score (0-56)								
Penicillamine or Trientine / day								
Zinc/day								
Video								
Picture of Kayser-Fleischer rings								

Global Assessment Scale for Wilson Disease: GAS for WD

GAS for WD Test Instructions

GAS for WD is a progressive two Tier disability scale; Tier 1: Global disability and Tier 2: Neurological Impairment. GAS for WD is administered to all WD patients at their first and subsequent visits

Tier 1: Global disability covers four domains: Liver (L), Cognition and behaviour (C), Motor (M) and Osseomuscular (O). Each domain is scored independently on an ascending six-point scale (0-5), generating four integers, each prefixed with its domain identifier (L, C, M and O, respectively). Individual scores are not summed as the domain scores reflect non-commensurable facets of the disease.

Tier 2: Neurological Impairment consists of 14 items, each scored on an ascending five-point scale (0-4). The individual scores are summed to obtain the Tier 2 total score (0-56).

- Grades based on historical accounts, such as Tier 1 (Cognition and behavior domain) and Tier 2 (Item 2: Scholastic performance; Item 3: Depression; Item 4: Psychosis) should reflect patient's clinical status over the previous **one month**. All other grades should reflect patient's clinical status at the time of administering the scale.
- Rate only "**what you see**". Grade each domain and item independently based upon the scale anchors, for instance, in a patient with incapacitating dystonia and tremor, grade both Tier 2 Item 5 (Dystonia) and Item 6 (Tremor), as 4.
- If a particular domain or item cannot be evaluated assign zero score appended by an asterisk (0*). For example, Tier 1 Osseomuscular domain is graded as 0* if there is no clinical evidence of osseomuscular involvement and X-rays are not available; similarly, Tier 2 Item 13 (KF rings) is graded as 0*, if KF rings are not visible with naked eye and a slit lamp examination cannot be done.
- If in doubt between two scores assign the higher score.
- Activities of Daily Living (ADL) refers to dressing, personal hygiene, eating, walking or any other day to day activity.

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GAS for WD Tier 1: Global disability

1. L - Liver^a

- L 0:** No past or ongoing / active liver disease
- L 1:** Liver disease in past but no ongoing /active liver disease
- L 2:** Ongoing / active liver disease but no evidence for cirrhosis ^b
- L 3:** Compensated cirrhosis
- L 4:** Decompensated ^c cirrhosis
- L 5:** Potentially life threatening liver disease^d

^a Clinical, biochemical and abdominal ultrasound evidence for liver disease; liver biopsy is optional.

^b Clinical or ultrasound evidence of cirrhosis or history of complications of cirrhosis; liver biopsy is optional.

^c Hepatic encephalopathy, porto-systemic bleeding, ascites.

^d Acute liver failure, liver disease with massive hemolysis or any liver disease requiring liver transplant.

2. C - Cognition and behavior^e

- C 0:** Normal
- C 1:** Symptoms noticed by parents, caregivers, immediate family members, or at school or work. Patient functions normally.
- C 2:** Obvious problems at home, school and work but can function at normal level with extra effort or help
- C 3:** Serious problems at home, school and work. Unable to function at normal level, *e.g.*, impaired interpersonal relationships, dropping grades at school or work
- C 4:** Unable to function independently except for simple ADL, *e.g.*, severely impaired interpersonal relations, discontinued school or work, needs considerable help, antidepressants or antipsychotics
- C 5:** Dependent on caregivers for even for simple ADL; institutionalized or needs to be restrained at home and is on antipsychotics or antidepressants

^e Related to intellectual decline, depression, psychosis

3. M - Motor^f

- M 0:** Asymptomatic or normal
- M 1:** Subtle clinical signs
- M 2:** Difficulty in ADL but independent
- M 3:** Requires help in ADL
- M 4:** Dependent on others for ADL
- M 5:** Bed bound

^f Neurological motor impairment

4. O - Osseomuscular^g

- O 0:** Normal
- O 1:** Abnormal skeletal X-ray; asymptomatic
- O 2:** Difficulty in ADL but independent
- O 3:** Requires help in ADL
- O 4:** Dependent on others for ADL
- O 5:** Fracture or bedbound

^g Bone, spinal or joint pain, swelling or deformity, or proximal muscle weakness

Tier 1 Score : L (0-5) _____ C (0-5) _____ M (0-5) _____ O (0-5) _____

GAS for WD Tier 2: Neurological impairment

<p>1. Wilson's facies</p> <ol style="list-style-type: none"> 0. Normal 1. Open mouth or facetious smile 2. Open mouth and facetious smile may have excessive salivation 3. Early dull look^h 4. Dull look^h <p>^hPsuedoptosis, decreased eye contact, decreased exploratory eye movements, drooping angle of mouth, delayed or no change in facial expressions</p>	<p>2. Scholastic Performance</p> <ol style="list-style-type: none"> 0. No intellectual declineⁱ; no change in academic or work performance 1. Mild intellectual declineⁱ; with help maintains grades or performance at work 2. Intellectual declineⁱ; clear deterioration in academic or work performance with dropping grades 3. Fails school and unable to continue schooling or work 4. Requires help in day to day intellectual activitiesⁱ <p>ⁱ Forgetfulness, difficulty in playing with friends, doing simple chores at home, running errands or difficulty in ADL (not explained by physical disability)</p>
<p>3. Depression ^j</p> <ol style="list-style-type: none"> 0. Absent 1. Subtle symptoms only recognized by parents, caregivers, immediate family members, or at school or work; not normal but does not interfere with family life, school or work 2. Mildly interferes with family life, school or work 3. Severely disrupts family life, school or work; requires antidepressants 4. Attempts suicide; requires hospitalization <p>^j Sadness of mood or disinterest in almost all or all activities with or without somatic symptoms (unexplained weight loss, alteration in sleep pattern, fatigue, loss of energy, feeling of worthlessness, inappropriate guilt, decreased concentration, indecisiveness)</p>	<p>4. Psychosis ^k</p> <ol style="list-style-type: none"> 0. Absent 1. Subtle symptoms; not normal but does not interfere with family life, school or work 2. Mildly interferes with family life, school or work 3. Severely disrupts family life, social life or relations and performance at school or work; requires antipsychotics 4. Attempted suicide or murder; requires restraining or hospitalization <p>^k Elevated mood, irritability, agitation, aggressiveness (e.g. throws temper tantrums, verbally or physically abusive or violent), flight of ideas, pressure of speech, motor restlessness (fidgety, hyperactive, runs away from home), impulsive behavior, hypersexuality, disinhibition, alcohol or drug abuse or addictions, delusions, hallucinations, threat or attempt of suicide or murder</p>
<p>5. Dystonia</p> <ol style="list-style-type: none"> 0. Absent 1. Dystonia that does not restrict any activity 2. Restricts ADL but independent 3. Restricts ADL; needs help 4. Bedbound 	<p>6. Tremor (rest, postural or action)</p> <ol style="list-style-type: none"> 0. Absent 1. Tremor that does not restrict any activity 2. Restricts ADL but independent 3. Restricts ADL; needs help 4. Bedbound
<p>7. Chorea</p> <ol style="list-style-type: none"> 0. Absent 1. Chorea that does not restrict any activity 2. Restricts ADL but independent 3. Restricts ADL; needs help 4. Bedbound 	<p>8. Parkinsonism</p> <ol style="list-style-type: none"> 0. Absent 1. Parkinsonism that does not restrict any activity 2. Restricts ADL but independent 3. Restricts ADL; needs help 4. Bedbound

<p>9. Speech</p> <ol style="list-style-type: none"> 0. Normal 1. Slurred but easily intelligible 2. Slurred; intelligible with difficulty 3. Unintelligible 4. Mute or Anarthric 	<p>10. Swallowing</p> <ol style="list-style-type: none"> 0. Normal 1. Chokes occasionally 2. Chokes frequently 3. Chokes with each meal 4. Aspiration pneumonia or on feeding tube
<p>11. Salivation</p> <ol style="list-style-type: none"> 0. Normal 1. Wets pillow at night; normal during day 2. Wet lips and angle of mouth; mouth filled with saliva; requires frequent wiping; 3. Intermittent drooling 4. Constant troublesome drooling 	<p>12. Posture and Gait (not due to solely osseomuscular involvement)</p> <ol style="list-style-type: none"> 0. Normal erect posture and gait 1. Abnormal posture but stands and walks independently. 2. Posture clearly abnormal; stands and walks with support of a walking stick or one person. 3. Cannot stand and walk without considerable help and can fall easily if unsupported. 4. Bedbound
<p>13. Kayser-Fleischer rings</p> <ol style="list-style-type: none"> 0. Absent 1. Visualized with slit lamp 2. Incomplete ring (restricted to superior, inferior or both corneal poles) visible with naked eye using torch 3. Complete thin ring visible with naked eye using torch 4. Complete thick ring visible with naked eye using torch 	<p>14. Uncommon Records the presence (1) or absence (0) of each of the following features:</p> <ul style="list-style-type: none"> • emotional lability • seizures over preceding 1 month • myoclonus • stereotypy • tics • pyramidal signs • eye movement abnormalities <p>Count the number of the above features that are present (upto a maximum of 4).</p>
<p>Tier 2 Total Score (0-56) _____</p>	

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