Chronic GVHD: Signs to Watch For

(Allogeneic Transplant Patients)
10 Signs of Chronic GVHD

1. Does my Skin feel tight or hard, have increased dryness, itching, or looks different (i.e., new rash, bumps, discoloration)?

2. Have I stopped sweating and have trouble cooling off (sweat glands)?

3. Am I losing hair (scalp or body) or are my nails brittle or ridged?

4. Are my wrists, fingers, or other joints stiff or painful?

5. Are my eyes dry, painful, or sensitive to wind or air conditioning?
6. Is my mouth dry or sensitive? Do things taste different? Do I have sores?

7. Do foods or pills get stuck when I swallow? (esophagus)?

8. Do I have trouble breathing, cough, or wheeze on exertion or at rest (lungs)?

9. Do I have vaginal dryness, pain, or itching? Do I have penile pain or pain when urinating (genitals)?

10. Have I lost weight without trying or can’t gain weight? (pancreatic or metabolic)
Skin Exam

Examine the entire body with a mirror or loved one’s help
Look for color and texture changes
Touch and pinch for change in texture and mobility:
- Thickened but moveable
- Thickened, moves poorly but can pinch
- Cannot pinch (hidebound)

Estimate percent of body area affected
Chronic GVHD Oral Exam
Where & What to Look For
If you see these changes, tell your doctor or LTFU
Oral Exam Tips

• Use a lamp or flashlight with a halogen light bulb

• Where to look?
  – lips, inside of cheeks, gums, tongue and throat

• What to look for?
  – redness
  – lacy patches
  – ulcers or sores
  – clear blisters on the back of the throat or inside of lips
Assess Joint stiffness
Report any suspected GVHD

• To your Doctor
  – Ask your doctor to fill out the GVHD assessment form and the range of motion form and fax it to LTFU at 206-667-5619

• To LTFU
  – If you have questions about whether your symptoms are chronic GVHD
  – If you are working with LTFU to adjust your medications
  – If possible, take digital pictures to show your signs and symptoms

• Always bring a list of all Medications/ Treatments when seeing your doctor or when contacting the LTFU office. You can email your medication/treatment list to LTFU at LTFU@seattlecca.org
**CHRONIC GRAFT-VERSUS-HOST DISEASE (GVHD) ASSESSMENT AND SCORING FORM**

<table>
<thead>
<tr>
<th>Patient: ___________________</th>
<th>Date of birth: ___________________</th>
<th>Date Evaluation: ___________________</th>
</tr>
</thead>
</table>

### PERFORMANCE SCORE:

<table>
<thead>
<tr>
<th>KPS</th>
<th>ECOG</th>
<th>LPS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**SCORING:**

- **SCORE 0:** Asymptomatic and fully active (ECOG 0; KPS or LPS 100%)
- **SCORE 1:** Symptomatic, fully ambulatory, restricted only in physically strenuous activity (ECOG 1, KPS or LPS 80-90%)
- **SCORE 2:** Symptomatic, ambulatory, capable of self-care, >50% of waking hours out of bed (ECOG 2, KPS or LPS 60-70%)
- **SCORE 3:** Symptomatic, limited self-care, >50% of waking hours in bed (ECOG 3-4, KPS or LPS <60%)

### SKIN†

**SCORE % BSA**

- **GVHD features to be scored by BSA:**
  - □ No BSA involved
  - □ 1-18% BSA
  - □ 19-50% BSA
  - □ >50% BSA

**Check all that apply:**

- □ Maculopapular rash/erythema
- □ Lichen planus-like features
- □ Sclerotic features
- □ Papulosquamous lesions or ichthyosis
- □ Keratosis pilaris-like GVHD

**SKIN FEATURES SCORE:**

- □ No sclerotic features
- □ Superficial sclerotic features “not hidebound” (able to pinch)

**Check all that apply:**

- □ Deep sclerotic features
- □ “Hidebound” (unable to pinch)
- □ Impaired mobility
- □ Ulceration

**Other skin GVHD features (NOT scored by BSA)**

**Check all that apply:**

- □ Hyperpigmentation
- □ Hypopigmentation
- □ Poikiloderma
- □ Severe or generalized pruritus
- □ Hair involvement
- □ Nail involvement
- □ Abnormality present but explained entirely by non-GVHD documented cause (specify): ________________________________
- □ Abnormality thought to represent GVHD PLUS other causes (specify): ________________________________

**MOUTH**

- □ No symptoms
- □ Mild symptoms **with** disease signs but not limiting oral intake
- □ Moderate symptoms **with** partial limitation of oral intake
- □ Severe symptoms with disease signs on examination **with** major limitation of oral intake

**Lichen planus-like features present:**

- □ Yes
- □ No

**Check all that apply:**

- □ Abnormality present but explained entirely by non-GVHD documented cause (specify): ________________________________
- □ Abnormality thought to represent GVHD PLUS other causes (specify): ________________________________

† Skin scoring should use both percentage of BSA involved by disease signs and the cutaneous features scales. When a discrepancy exists between the percentage of total body surface (BSA) score and the skin feature score, OR if superficial sclerotic features are present (Score 2), but there is impaired mobility or ulceration (Score 3), the higher level should be used for the final skin scoring.
<table>
<thead>
<tr>
<th>EYES</th>
<th>SCORE 0</th>
<th>SCORE 1</th>
<th>SCORE 2</th>
<th>SCORE 3</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No symptoms</td>
<td>Mild dry eye symptoms not affecting ADL (requirement of lubricant eye drops ≤ 3 x per day)</td>
<td>Moderate dry eye symptoms partially affecting ADL (requiring lubricant eye drops &gt; 3 x per day or punctal plugs), WITHOUT new vision impairment due to KCS</td>
<td>Severe dry eye symptoms significantly affecting ADL (special eyewear to relieve pain) OR unable to work because of ocular symptoms OR loss of vision due to KCS</td>
</tr>
</tbody>
</table>

Keratoconjunctivitis sicca (KCS) confirmed by ophthalmologist:
- Yes
- No
- Not examined

☐ Abnormality present but explained entirely by non-GVHD documented cause (specify): ____________________________________________
☐ Abnormality thought to represent GVHD PLUS other causes (specify) :

<table>
<thead>
<tr>
<th>GI Tract</th>
<th>Check all that apply:</th>
<th>SCORE 0</th>
<th>SCORE 1</th>
<th>SCORE 2</th>
<th>SCORE 3</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No symptoms</td>
<td>Symptoms without significant weight loss* (&lt;5%)</td>
<td>Symptoms associated with mild to moderate weight loss* (5-15%) OR moderate diarrhea without significant interference with daily living</td>
<td>Symptoms associated with significant weight loss* &gt;15%, requires nutritional supplement for most calorie needs OR esophageal dilation OR severe diarrhea with significant interference with daily living</td>
<td></td>
</tr>
</tbody>
</table>

☐ Abnormality present but explained entirely by non-GVHD documented cause (specify): ____________________________________________
☐ Abnormality thought to represent GVHD PLUS other causes (specify) :

| LIVER | Normal total bilirubin and ALT or AP < 3 x ULN | Normal total bilirubin with ALT ≥ 3 to 5 x ULN or AP ≥ 3 x ULN | Elevated total bilirubin but ≤3 mg/dL or ALT > 5 ULN | Elevated total bilirubin > 3 mg/dL |

☐ Abnormality present but explained entirely by non-GVHD documented cause (specify): ____________________________________________
☐ Abnormality thought to represent GVHD PLUS other causes (specify) :

| LUNGS** | Symptom score: | No symptoms | Mild symptoms (shortness of breath after climbing one flight of steps) | Moderate symptoms (shortness of breath after walking on flat ground) | Severe symptoms (shortness of breath at rest; requiring O2) |

☐ Lung score: FEV1 ≥80% | FEV1 60-79% | FEV1 40-59% | FEV1 <39% |

Pulmonary function tests
- Not performed

☐ Abnormality present but explained entirely by non-GVHD documented cause (specify): ____________________________________________
☐ Abnormality thought to represent GVHD PLUS other causes (specify) :

* Weight loss within 3 months. **Lung scoring should be performed using both the symptoms and FEV1 scores whenever possible. **FEV1 should be used in the final lung scoring where there is discrepancy between symptoms and FEV1 scores.
CHRONIC GRAFT-VERSUS-HOST DISEASE (GVHD) ASSESSMENT AND SCORING FORM

Patient:________________________________________________________________________________________

<table>
<thead>
<tr>
<th>JOINTS AND FASCIA</th>
<th>SCORE 0</th>
<th>SCORE 1</th>
<th>SCORE 2</th>
<th>SCORE 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>P-ROM score</td>
<td>□ No symptoms</td>
<td>□ Mild tightness of arms or legs, normal or mild decreased range of motion (ROM) AND not affecting ADL</td>
<td>□ Tightness of arms or legs OR joint contractures, erythema thought due to fasciitis, moderate decrease ROM AND mild to moderate limitation of ADL</td>
<td>□ Contractures WITH significant decrease of ROM AND significant limitation of ADL (unable to tie shoes, button shirts, dress self etc.)</td>
</tr>
<tr>
<td>Shoulder (1-7):___</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Elbow (1-7):_____</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wrist/finger (1-7):___</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ankle (1-4):____</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

□ Abnormality present but explained entirely by non-GVHD documented cause (specify): _____________________________________________________________

□ Abnormality thought to represent GVHD PLUS other causes (specify): ________________________________________________________________

<table>
<thead>
<tr>
<th>GENITAL TRACT</th>
<th>SCORE 0</th>
<th>SCORE 1</th>
<th>SCORE 2</th>
<th>SCORE 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>(See Supplemental figure‡)</td>
<td>□ No signs</td>
<td>□ Mild signs‡ and females with or without discomfort on exam</td>
<td>□ Moderate signs‡ and may have symptoms with discomfort on exam</td>
<td>□ Severe signs‡ with or without symptoms</td>
</tr>
<tr>
<td>Currently sexually active</td>
<td>□ Yes</td>
<td>□ No</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

□ Abnormality present but explained entirely by non-GVHD documented cause (specify): _____________________________________________________________

□ Abnormality thought to represent GVHD PLUS other causes (specify): ________________________________________________________________

Other indicators, clinical features or complications related to chronic GVHD (check all that apply and assign a score to severity (0-3) based on functional impact where applicable none – 0, mild -1, moderate -2, severe – 3)

□ Ascites (serositis)___ | □ Myasthenia Gravis___
□ Pericardial Effusion___ | □ Peripheral Neuropathy___ | □ Eosinophilia > 500/μl___
□ Pleural Effusion(s)___ | □ Polymyositis___ | □ Platelets <100,000/μl ___
□ Nephrotic syndrome___ | □ Weight loss>5%* without GI symptoms___ | □ Others (specify):___________

Biopsy obtained: □ Yes □ No Organ biopsied:______________ GVHD confirmed by histology: □ Yes □ No

Overall GVHD Severity
(Reevaluation by the provider)

□ No GVHD □ Mild □ Moderate □ Severe

Change from prior evaluations:
□ No prior or current GVHD □ Improved □ Stable □ Worse □ N/A (baseline)

Photographic Range of Motion (P-ROM):

Completed by:_________________________________________________________ Date form completed:__________________________
Name_________________________________________ DOB___/___/___  Assessment Date: ___/___/____

Circle the number that best matches how flexible you are in each of these positions

<table>
<thead>
<tr>
<th>Minimum</th>
<th>Flexibility</th>
<th>Maximum</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Shoulder</td>
<td>7</td>
</tr>
<tr>
<td>1</td>
<td>Elbow</td>
<td>7</td>
</tr>
<tr>
<td>1</td>
<td>Wrist and fingers</td>
<td>7</td>
</tr>
<tr>
<td>1</td>
<td>Foot Dorsiflexion</td>
<td>4</td>
</tr>
</tbody>
</table>

Please ask your doctor to complete and Fax or email this form to the LTFU Office at (800)376-8197 or LTFU@seattlecca.org
**Supplement Figure – Genital Tract Chronic Graft-versus-Host Assessment and Scoring Form**

<table>
<thead>
<tr>
<th>Name:______________________________</th>
<th>Date of birth:___________</th>
<th>Assessment date:_________</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>GENITAL TRACT</th>
<th>SCORE 0</th>
<th>SCORE 1</th>
<th>SCORE 2</th>
<th>SCORE 3</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No signs</td>
<td>Mild signs and females may have symptoms* WITH discomfort on exam</td>
<td>Moderate signs and may have symptoms* with discomfort on exam</td>
<td>Severe signs with or without symptoms *</td>
</tr>
</tbody>
</table>

**Check:**
- [ ] Male
- [ ] Female

**Currently sexually active:**
- [ ] Yes
- [ ] No

**Check all signs that apply:**
- [ ] Lichen Planus-like features
- [ ] Lichen sclerosis-like features
- [ ] Vaginal scarring (female)
- [ ] Clitoral/labial agglutination (female)
- [ ] Labial resorption (female)
- [ ] Erosions
- [ ] Fissures
- [ ] Ulcers
- [ ] Phimosis (male)
- [ ] Urethral meatus scarring/stenosis (male)

- [ ] Abnormality present but NOT thought to represent GVHD (specify cause): ________________________________
- [ ] Abnormality thought to represent GVHD PLUS other causes (specify cause): ________________________________

*Genital symptoms are not specific to cGVHD and can represent premature gonadal failure or genital tract infection.

If a gynecologist is unavailable, external examination may be performed to determine “discomfort on exam” as follows:

a) Spread the labia majora to inspect the vulva for the above signs. Touch the vestibular gland openings (Skene’s and Bartholin’s), labia minora and majora gently with a qtip. Vulvar pain elicited by the gentle touch of a qtip is classified as discomfort on examination. Palpate the vaginal walls with a single digit to detect bands, shortening, narrowing or other signs of vaginal scarring.

b) If the woman is sexually active, determine whether qtip palpation or gentle palpation of scarred ridges elicits pain similar to that which the woman experiences during intercourse.

**Female genitalia:** Severity of signs:
1) Mild (any of the following): erythema on vulvar mucosal surfaces, vulvar lichen-planus or vulvar lichen-sclerosis.
2) Moderate (any of the following): erosive inflammatory changes of the vulvar mucosa, fissures in vulvar folds
3) Severe (any of the following): labial fusion, clitoral hood agglutination, fibrinous vaginal adhesions, circumferential fibrous vaginal banding, vaginal shortening, synechia, dense sclerotic changes, and complete vaginal stenosis.

**Male genitalia:** Diagnostic features include lichen planus-like or lichen sclerosis-like features and phimosis or urethral scarring or stenosis. Severity of signs:
1) Mild: lichen planus-like feature;
2) Moderate: lichen sclerosus-like feature or moderate erythema;
3) Severe: phimosis or urethral/meatal scarring.

**Biopsy obtained:**
- [ ] Yes
- [ ] No

Site biopsied: ________________

GVHD confirmed by histology:
- [ ] Yes
- [ ] No

**Change from previous evaluation:**
- [ ] No prior or current GVHD
- [ ] Improved
- [ ] Stable
- [ ] Worse
- [ ] N/A (baseline)

Completed by (print name): ________________________________ Date form completed: __________