

**Pediatric adaption of NIH 2014 cGVHD diagnosis/staging for clinical practice  
German-Austrian-Swiss GVHD Consortium**

completed by:

date

patient name

▶ please score/check the worst manifestation

▶ diagnostic features are marked **bold**

**classification:**

- feat. of acute GVHD
- feat. of classic cGVHD
- both

**onset type at diagnosis:**

- de novo
- quiescent
- progressive

height: \_\_\_\_\_

weight: \_\_\_\_\_

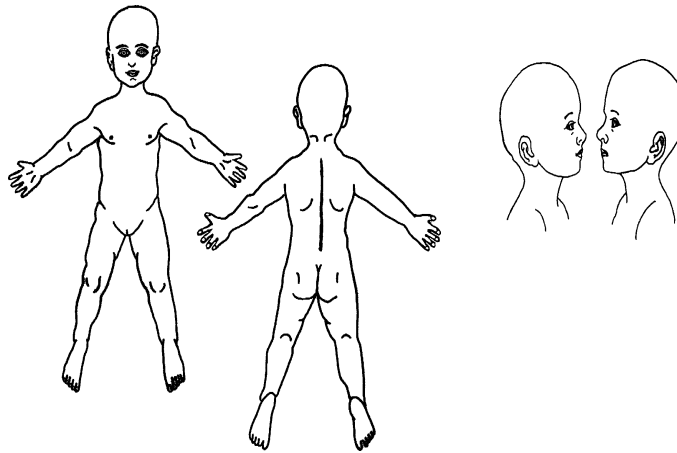
symptoms/features	Score 0	Score 1	Score 2	Score 3
<b>KPS/LPS:</b> %	<input type="checkbox"/> asymptomatic and fully active (KPS/LPS 100%)	<input type="checkbox"/> sympt., fully amb., restricted only in physically strenuous activity (KPS/LPS 80-90%)	<input type="checkbox"/> sympt., amb., capable of self-care, >50% of waking hours out of bed (KPS/LPS 60-70%)	<input type="checkbox"/> sympt., limited self-care >50% of waking hours in bed (KPS/LPS < 60%)

**SKIN**

Feat. scored by BSA:	no BSA involved	1-18% BSA	19-50% BSA	> 50% BSA
<input type="checkbox"/> maculopapular rash/erythema	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> <b>lichen planus-like features</b>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> <b>sclerotic features:</b>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> <b>lichen sclerosus-like</b>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> papulosquamous lesions	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> ichthyosis	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> keratosis pilaris-like GVHD	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Feat. not scored by BSA:

- hyperpigmentation
- hypopigmentation/ depigmentation
- poikiloderma**
- severe pruritus
- hair involvement
- nail involvement
- sweat impairment
- abnormality present but explained entirely by non-GVHD cause (specify):
- abnormality thought to represent GVHD **PLUS** other causes (specify):



%BSA:  
 child: head front/back 9 / 9  
 back 18, chest 18,  
 arm left 9, arm right 9  
 leg left 13,5, leg right 13,5  
 adult: head front/back 4,5 / 4,5  
 back 18, chest 18  
 arm left 9, arm right 9  
 leg left 18, leg right 18  
 palm: 1,5

<b>sclerotic features:</b>	<input type="checkbox"/> no sclerotic features	<input type="checkbox"/> superficial sclerotic features "not hidebound" (able to pinch)	<input type="checkbox"/> deep sclerotic features "hidebound" (unable to pinch) <input type="checkbox"/> impaired mobility <input type="checkbox"/> ulceration
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**MOUTH**

	no symptoms	mild sympt with disease signs but not limiting oral intake significantly	moderate sympt. with disease signs with partial limitation of oral intake	severe sympt. with disease signs on examination with major limitation of oral intake
<input type="checkbox"/> erythema	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> <b>lichen planus-like features</b>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> hyperkeratot. plaques	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> mucocelles <input type="checkbox"/> pseudomembranes	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> ulcers <input type="checkbox"/> mucosal atrophy	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> dryness <input type="checkbox"/> pain	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify):				
<input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				

symptoms/features	Score 0	Score 1	Score 2	Score 3
<b>EYES</b>				
<input type="checkbox"/> keratokonjunktivitis sicca (KCS) <input type="checkbox"/> confirmed by ophthalmologist <input type="checkbox"/> dryness <input type="checkbox"/> pain <input type="checkbox"/> photophobia <input type="checkbox"/> blepharitis <input type="checkbox"/> pseudomembranes <input type="checkbox"/> ulcers	<input type="checkbox"/> no symptoms	<input type="checkbox"/> mild dry eye sympt. not affecting ADL (requirement of lubricant eye drops ≤ 3 x per day)	<input type="checkbox"/> moderate dry eye sympt. partially affecting ADL (lubricant eye drops >3 x/d or punctual plugs) without new vision impairment due to KCS	<input type="checkbox"/> severe dry eye sympt. significantly affecting ADL (special eyewear to relieve pain) or unable to work because of ocular sympt or loss of vision due to KCS
<input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify): <input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				
<b>GI TRACT</b>				
<input type="checkbox"/> esophageal web/ <b>prox stricture or ring</b> <input type="checkbox"/> dysphagia <input type="checkbox"/> abdominal pain <input type="checkbox"/> anorexia <input type="checkbox"/> failure to thrive <input type="checkbox"/> nausea <input type="checkbox"/> vomiting <input type="checkbox"/> diarrhea <input type="checkbox"/> weight loss ≥ 5%	<input type="checkbox"/> no symptoms	<input type="checkbox"/> symptoms without significant weight loss (5%)	<input type="checkbox"/> sympt. associated with mild to moderate weight loss (5-15%) or moderate diarrhea without significant interference with daily living	<input type="checkbox"/> symptoms associated with significant weight loss (> 15%) requires nutritional supplement for most calorie needs or esophageal dilatation or severe diarrhea with signif. Interference with daily living
<input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify): <input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				
<b>LIVER</b>				
<input type="checkbox"/> hepatitic pattern	<input type="checkbox"/> normal total bili and ALT or AP < 3 ULN	normal total bili <input type="checkbox"/> with ALT ≥ 3-5x ULN <input type="checkbox"/> or AP ≥ 3 x ULN	elevated total bili <input type="checkbox"/> but ≤ 3 mg/dl or <input type="checkbox"/> ALT > 5 ULN	<input type="checkbox"/> elevated total bili > 3 mg/dl
<input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify): <input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				
<b>LUNGS</b>				
<b>FEV1:</b> _____% <b>MEF25:</b> _____% <b>FVC:</b> _____% <b>MEF50:</b> _____% <b>DLCO:</b> _____% <b>MEF75:</b> _____% <b>RV:</b> _____ <input type="checkbox"/> RV/TLC > 120% <b>CT:</b> _____	<input type="checkbox"/> no symptoms <input type="checkbox"/> FEV1 ≥ 80%	<input type="checkbox"/> mild symptoms (shortness of breath after climbing one flight of steps) <input type="checkbox"/> FEV1 60-79%	<input type="checkbox"/> moderate symptoms (shortness of breath after walking on flat ground) <input type="checkbox"/> FEV1 40-59%	<input type="checkbox"/> severe symptoms (shortness of breath at rest; requiring O2) <input type="checkbox"/> FEV1 ≤ 39%
<input type="checkbox"/> pulmonary function test not performed <input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify): <input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				
<b>JOINTS AND FASCIA</b>				
<input type="checkbox"/> ped P-ROM score (see below) <input type="checkbox"/> edema <input type="checkbox"/> fasciitis <input type="checkbox"/> muscle cramps <input type="checkbox"/> athralgia	<input type="checkbox"/> no symptoms	<input type="checkbox"/> mild tightness, normal or mild ↓ of range of motion (ROM) not affecting ADL	<input type="checkbox"/> tightness or joint contractures, fasciitis, moderate ↓ of ROM, mild - moderate ↓ of ADL	<input type="checkbox"/> contractures, fasciitis significant ↓ of ROM, significant ↓ of ADL
<input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify): <input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				
<b>GENITAL TRACT</b>				
<input type="checkbox"/> erosions, fissures <input type="checkbox"/> lichen planus-like features <input type="checkbox"/> lichen sclerosus-like features <input type="checkbox"/> labial/ vaginal scarring <input type="checkbox"/> phimosis	<input type="checkbox"/> no signs	<input type="checkbox"/> mild signs	<input type="checkbox"/> moderate signs	<input type="checkbox"/> severe signs with or without symptoms
<input type="checkbox"/> abnormality present but explained entirely by non-GVHD cause (specify): <input type="checkbox"/> abnormality thought to represent GVHD <b>PLUS</b> other causes (specify):				

**Overall GVHD severity**

<input type="checkbox"/> no cGVHD		maximum individual score
<input type="checkbox"/> mild:	max. score of 1 in any affected organ, max. 2 organs affected, no lung involvement	total score (sum)
<input type="checkbox"/> moderate:	≥3 organ with max score 1 or max. score of 2 in any affected organ, lung score max 1	average score (total score/24)
<input type="checkbox"/> severe:	score 3 in any affected organ, lung score 2-3	number of affected organs

Other indicators, clinical features or complications related to cGVHD	biopsy:	Yes <input type="checkbox"/> No <input type="checkbox"/>
check all that apply and assign a severity score (0-3) based on functional impact	date:	
<input type="checkbox"/> ascites (serositis)	organ:	
<input type="checkbox"/> pericardial effusion	GVHD confirmed? Yes <input type="checkbox"/> No <input type="checkbox"/>	
<input type="checkbox"/> pleural effusion		suspicion
<input type="checkbox"/> nephrotic syndrome		
<input type="checkbox"/> others (specify)		

**Intensity of current immunosuppression**

- None
- Mild (single agent prednisone < 0.5 mg/kg/day)
- Moderate (prednisone ≥ 0.5 mg/kg/day and/or any single agent/modality)
- High (2 or more agents/modalities ± prednisone ≥ 0.5 mg/kg/day)

**Therapeutic intent at the time of clinic visit**

- Decision to decrease systemic therapy because cGVHD is better
- Decision is to not change current systemic therapy because cGVHD is stable
- Decision is to increase systemic therapy because cGVHD is worse
- Alter systemic therapy due to its toxicity
- Substitute systemic therapy due to lack of response
- Withdraw systemic therapy due to lack of response
- Not applicable

**Clinician's impression of activity**

- Inactive, off systemic therapy or topical immunosuppression
- Inactive, on systemic therapy or topical immunosuppression
- Active, irrespective of the level of current therapy
- Highly active, irrespective of the level of current therapy

**Pediatric Photographic Range of Motion (adapted ped P-ROM)**

please mark appropriate number

shoulder: 1 (worst) 2 3 4 5 (normal)



elbow: 1 (worst) 2 3 4 (normal)



wrist / finger: 1 (worst) 2 3 4 (normal)



global flexion: 1 (worst) 2 3 4 (normal)



ankle: 1 (worst) 2 3 (normal)

