

# **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	Score1	Score 2	Score 3
KPS/LPS: %	<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):

%BSA:

child:

adult:

palm: 1,5

sclerotic features:	<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				
<input type="checkbox"/> erythema	<input type="checkbox"/> no symptoms	<input type="checkbox"/> mild sympt with disease signs but not limiting oral intake significantly	<input type="checkbox"/> moderate sympt. with disease signs with partial limitation of oral intake	<input type="checkbox"/> severe sympt. with disease signs on examination with major limitation of oral intake
<input type="checkbox"/> <b>lichen planus-like features</b>				
<input type="checkbox"/> hyperkeratot. plaques				
<input type="checkbox"/> mucocoeles <input type="checkbox"/> pseudomembranes				
<input type="checkbox"/> ulcers <input type="checkbox"/> mucosal atrophy				
<input type="checkbox"/> dryness <input type="checkbox"/> pain				
<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/ prox stricture or ring <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"/> hepatic 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	<input type="checkbox"/> elevated total bili but ≤ 3 mg/dl or 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"/> currently sexually active      coitarche: _____ years				
<input type="checkbox"/> hormonal status: hypogonadism <input type="checkbox"/> signs of vaginal hypoestrogenization				
<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

check all that apply and assign a severity score (0-3) based on functional impact			biopsy:	Yes <input type="checkbox"/> No <input type="checkbox"/>
<input type="checkbox"/> ascites (serositis)	<input type="checkbox"/> myasthenia gravis	<input type="checkbox"/> diabetes	date:	
<input type="checkbox"/> pericardial effusion	<input type="checkbox"/> peripheral nervous manifestations	<input type="checkbox"/> eosinophilia >500 /ul	organ:	
<input type="checkbox"/> pleural effusion	<input type="checkbox"/> central nervous manifestations	<input type="checkbox"/> platelets <100 000/ul	GVHD confirmed? Yes <input type="checkbox"/> No <input type="checkbox"/>	
<input type="checkbox"/> nephrotic syndrome	<input type="checkbox"/> polymyositis	<input type="checkbox"/> hypo/hyperglobulinemia		suspicion
<input type="checkbox"/> others (specify)	<input type="checkbox"/> weight loss >5% without GI symptoms	<input type="checkbox"/> auto-antibodies		
		<input type="checkbox"/> immune thyroiditis		

### Change from prior evaluation:

☐ improved ☐ stable ☐ worse ☐ comment: \_\_\_\_\_

### 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)



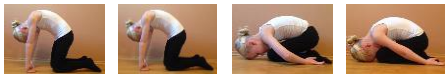
elbow: 1 (worst) 2 3 4 (normal)



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



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



ankle: 1 (worst) 2 3 (normal)

