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

German-Austrian-Swiss (GVHD Consortiun	n			
completed by:		date	patient name		
▶ please score/check the worst mani	festation	classification:	onset type at diagnosis:		
▶ diagnostic features are marked bo	ld	□ feat. of acute GVHD	□ de novo	height:	
		□ feat.of classic cGVHD	□ quiescent		
		□ both	□ progressive	weight:	
symptoms/features	Score 0	Score1	Score 2	Score 3	
KPS/LPS: %	□ asymptomatic and fully active (KPS/LPS 100%)	□ sympt., fully amb., restricted only in physically strenous activity (KPS/LPS 80-90%)	□ sympt.,amb., capable of self-care, >50% of waking hours out of bed (KPS/LPS 60-70%)	□ 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	
□ maculopapular rash/erythema					
□ lichen planus-like features					
□ sclerotic features:					
□ lichen sclerosus-like					
□ papulosquamous lesions					
□ ichthyosis					
□ keratosis pilaris-like GVHD					
Feat. not scored by BSA:					
□ hyperpigmentation	(33)			%BSA:	
□ hypopigmentation/ depigmentation	Š			child: head front/back 9 / 9	
□ poikiloderma		\ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \ \		back 18, chest 18, arm left 9, arm right 9	
□ severe pruritus	/ / /			leg left 13,5, leg right 13,5 adult: head front/back 4,5 / 4,5	
□ hair involvement			$)$ γ γ $($	back 18, chest 18 arm left 9, arm right 9	
□ nail involvement	\sim	2/1/1/3	/ / / / /	leg left 18, leg right 18	
□ sweat impairment	/ /\ \		'	palm: 1,5	
□ abnormality present but explained	/*/ \\\	141			
entirely by non-GVHD cause (specify):	1/\\]	/~/ _\			
		1 // //			
= abnormality thought to represent	₩ ₩	\mathcal{H}			
□ abnormality thought to represent GVHD PLUS other causes (specify):		and both			
GVIID FLOS other causes (specify).					
sclerotic features:	□ no sclerotic		□ superficial	□ deep sclerotic features	
	features		sclerotic features "not hidebound" (able to pinch)	□ "hidebound" (unable to pinch)□ impaired mobility□ ulceration	
MOUTH					
□ erythema	□ no symptoms	□ mild sympt with	□ moderate sympt.	□ severe sympt. with	
□ lichen planus-like features		disease signs but	with disease signs	disease signs on examination	
□ hyperkeratot. plaques		not limiting oral	with partial limitation	with major limitation	
□ mucoceles □ pseudomembranes		intake significantly	of oral intake	of oral intake	
□ ulcers □ mucosal atrophy					
□ dryness □ pain					

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

EYES	a keratokonjunktivitis s a confirmed by opthalr a dryness			Score1	Score 2	Score 3
continend by optimized popularizations of the partial particulary affecting ADL (u)brocart eye drops (particularly affecting ADL (partially affecting ADL (particular particular	confirmed by opthali					
dyness pain (requirement of (lubnoant eye drops 23 xid or punctual plugh) unable to work because of season production 24 x per day) without new vision symptor loss of vision due to KC state of the production 25 x per day) without new vision symptor loss of vision due to KC state of the production 25 x per day) without new vision symptor loss of vision due to KC state of the production 25 x per day) without new vision symptor loss of vision due to KC state of the production 25 x per day) symptor 25 x per day symptor 25 x per day) symptor	dryness	sicca (KCS)	□ no symptoms	□ mild dry eye sympt.	□ moderate dry eye sympt.	□ severe dry eye sympt.
dryness pain (requirement of (lubricant eye drops (special syeware to relieve pain) hotophotophoto bispharetts bis	dryness		, ,			
photophobia is beyinariis Libricant eye drops 2-3 x0 or punctual plugs unable to work because of oculi passocinembranes Unifors 2-3 x per day) written the every window symptom sympt				_		
special contents and the special contents of the speci	p	•				
## Benomality present but explained enterly by non-GVHD cause (specify): ## abnormality throught to represent GVHD PLUS other causes (specify): ## approximation of the properties of the proximation of t	pseudomembranes -				without new vision	sympt or loss of vision due to KCS
esphageal web/ prox stricture or ring dispribagia		· ·			impairement due to NOO	
esphageal web/ prox stricture or ring dispribagia	GLTRACT					
significant weight loss (> 15%) weight loss (> 15%) requires nutrional supplement for anorexals in abdominal pain anorexals in failure to thrive nausea requires nutrional supplement for anorexals requires require			□ no symptoms	□ symptoms without	□ sympt. associated with	□ symptoms associated with
dysphagia abdominal pain loss (5%) weight toss (5-15%) requires nutritional supplement from a morexal a failure to thrive nutries of the provided diarrha and the provided	-	ina	,,			• •
anorexia of failure to thrive in ausea of vomiting without significant exphageal dilatation or supplied and received to the property of the pr	•	•				
nausea community working without significant community c		•		(****)	. ,	
interference with severe diarrhea with daily living signif. Interference with daily living signi						
daily living signif. Interference with daily living abnormality present but explained entirely by non-GVHD cause (specify): a bnormality thought to represent GVHD PLUS other causes (specify): a bnormality thought to represent GVHD PLUS other causes (specify): and ALT or AP		ŭ			-	
abnormality present but explained entirely by non-GVHD cause (specify): abnormality thought to represent GVHD PLUS other causes (specify):		0.9 1000 £ 0 /0				
normal total bili normal bili n		to represent GVH	ID PLUS other causes (sp	ecify):		
and ALT or AP			T			
< 3 ULN	hepatitic pattern					□ elevated total bili > 3 mg/dl
abnormality present but explained entirely by non-GVHD cause (specify): abnormality thought to represent GVHD PLUS other causes (specify): LUNGS			and ALT or AP	□ with ALT ≥ 3-5x ULN	□ but ≤ 3 mg/dl or	
LUNGS FEV1:			< 3 ULN	□ OF AP ≥ 3 X ULN	□ ALI > 5 ULN	
EUNGS FEV1:%		· ·				
FEV1:% MEF25:% no symptoms mild symptoms moderate symptoms severe symptoms FVC:% MEF50:% FEV1 ≥ 80% (shortness of breath (shor	abnormanty thought	to represent GVII	ib i Loo offici cadaca (apr	50ny).		
FVC:% MEF50:%	LUNGS					
A green process of the part of the process of the process of the process of the part of t	FEV1:% N	/IEF25:%	□ no symptoms	□ mild symptoms	□ moderate symptoms	□ severe symptoms
A green process of the part of the process of the process of the process of the part of t	FVC:% N	MEF50:%	□FEV1 ≥ 80%	(shortness of breath	(shortness of breath	(shortness of breath at rest;
RV:		· <u> </u>				
CT:				_	_	
pulmonary function test not performed abnormality present but explained entirely by non-GVHD cause (specify): abnormality thought to represent GVHD PLUS other causes (specify): DOINTS AND FASCIA						
abnormality present but explained entirely by non-GVHD cause (specify): abnormality thought to represent GVHD PLUS other causes (specify): JOINTS AND FASCIA ped P-ROM score (see below)		est not performed	<u> </u>	2.21.00.0%	2.27.1000%	
JOINTS AND FASCIA ped P-ROM score (see below)	'	•		(specify):		
ped P-ROM score (see below)	abnormality thought	to represent GVH	ID PLUS other causes (sp	ecify):		
ped P-ROM score (see below)	IOINTS AND	FACCIA				
normal or mild \(\) of contractures, fasciitis, significant \(\) of ROM, significant \(\) of ADL not affecting ADL mild - moderate \(\) of ADL abnormality present but explained entirely by non-GVHD cause (specify): abnormality thought to represent GVHD PLUS other causes (specify): GENITAL TRACT erosions, fissures no signs mild signs moderate signs severe signs with or without symptoms lichen planus-like features lichen sclerosus-like features labial/ vaginal scarring phimosis currently sexually active coitarche:years	JOIN 15 AND				Alaska and a last at	
range of motion (ROM) moderate \ of ROM, significant \ of ADL abnormality present but explained entirely by non-GVHD cause (specify): abnormality thought to represent GVHD PLUS other causes (specify): GENITAL TRACT a erosions, fissures no signs mild signs moderate signs severe signs with or without symptoms blichen planus-like features lichen sclerosus-like features labial/ vaginal scarring phimosis currently sexually active coltarche:years	LD DOM /	•	□ no symptoms	_		,
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GENITAL TRACT cerosions, fissures no signs no mild signs no moderate signs severe signs with or without symptoms clichen planus-like features symptoms clichen sclerosus-like features phimosis currently sexually active coitarche:years	edema =	aumaigia		not affecting ADL	mild - moderate ↓ of ADL	
erosions, fissures	a edema a muscle cramps	but explained enti		(specify):	mild - moderate ↓ of ADL	
symptoms a lichen planus-like features a labial/ vaginal scarring phimosis currently sexually active coitarche:years	a edema a muscle cramps	but explained enti		(specify):	mild - moderate ↓ of ADL	
lichen sclerosus-like features labial/ vaginal scarring phimosis currently sexually active coitarche:years	edema muscle cramps abnormality present abnormality thought	but explained enti to represent GVH		(specify):	mild - moderate ↓ of ADL	
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currently sexually active coitarche:years	a dedema and muscle cramps and abnormality present abnormality thought GENITAL TR a erosions, fissures	but explained enti to represent GVH	ID PLUS other causes (sp	(specify): ecify):		
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hormonal status: hypogonadism signs of vaginal hypoestrogenization	dedema de	but explained entito represent GVH ACT eatures	ID PLUS other causes (sp	(specify): ecify):		
	abnormality present abnormality thought GENITAL TR perosions, fissures lichen planus-like for lichen sclerosus-like labial/ vaginal scar	but explained entito represent GVH ACT reatures re features ring phimosis	ID PLUS other causes (sp	(specify): ecify):		

Overall GVHD severity maximum individual score □ no cGVHD max. score of 1 in any affected organ, max. 2 organs affected, no lung involvement total score (sum) □ mild: ≥3 organ with max score 1 or max. score of 2 in any affected organ, lung score max 1 average score (total score/24) □ moderate: score 3 in any affected organ, lung score 2-3 number of affected organs □ severe: Other indicators, clinical features or complications related to cGVHD biopsy: check all that apply and assign a severity score (0-3) based on functional impact date: □ ascites (serositis) □ myasthenia gravis □ diabetes organ: □ pericardial effusion □ peripheral nervous manifestations □ eosinophilia >500 /ul □ pleural effusion GVHD confirmed? Yes □ No □ □ central nervous manifestations □ platelets <100 000/ul □ nephrotic syndrome □ polymyositis □ hypo/hyperglobulinemia □ others (specify) □ weight loss >5% without GI symptoms auto-antibodies □ 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 $\hfill \square$ Decision is to not change current systemic therapy because cGVHD □ 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 Yes □ No □

suspicion

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

please markappropriate number

shoulder:	1 (worst)	2	3	4	5 (normal)
		1			
ellbow:	1 (worst)	2	3	4 (normal)	
		4		-	
wrist / finger:	1 (worst)	2	3	4 (normal)	
global flexion:	1 (worst)	2	3	4 (normal)	
	1	1		R	
ankle:	1 (worst)	2	3 (normal)		
		L			