新規申請時の初期治療（平成　　年　　月）または直近の急性増悪時治療（平成　　年　　月）			治療効果
内服治療	エトレチナート	1.あり（　　）mg/日　　2.なし	1.あり　2.なし　3.不明
	シクロスポリンMEPC	1.あり（　　）mg/kg/日　　2.なし	1.あり　2.なし　3.不明
	メトトレキセート	1.あり（　　）mg/週　　2.なし	1.あり　2.なし　3.不明
	副腎皮質ステロイド	1.あり（プレドニゾロン換算　　mg/日）　　2.なし	1.あり　2.なし　3.不明
	その他	1.あり（内容　　処方量　　）　2.なし	1.あり　2.なし　3.不明
生物学的製剤	インフリキシマブ	1.あり（　　）mg/日　使用週（0,　,　,　週）（　　週毎） 2.なし	1.あり　2.なし　3.不明
	（複数回答可→）	[使用目的：1.皮膚病変　2.関節炎　3.その他（　　）]	
	その他	1.あり（薬剤名：　　）（用量:　　用法:　　） 2.なし	1.あり　2.なし　3.不明
	（複数回答可→）	[使用目的：1.皮膚病変　2.関節炎　3.その他（　　）]	
外用・光線療法等	副腎皮質ステロイド外用	1.あり　　2.なし	1.あり　2.なし　3.不明
	活性型ビタミンD3外用	1.あり　　2.なし	1.あり　2.なし　3.不明
	光線療法	1.あり[1.PUVA　2.NB-UVB　3.その他（　　）]　2.なし	1.あり　2.なし　3.不明
	その他	1.あり（内容　　量　　）　2.なし	1.あり　2.なし　3.不明
主たる維持療法		治療（最近１年以内の状況）	治療効果
内服治療	エトレチナート	1.あり（　　）mg/日　　2.なし	1.あり　2.なし　3.不明
	シクロスポリンMEPC	1.あり（　　）mg/kg/日　　2.なし	1.あり　2.なし　3.不明
	メトトレキセート	1.あり（　　）mg/週　　2.なし	1.あり　2.なし　3.不明
	副腎皮質ステロイド	1.あり（プレドニゾロン換算　　mg/日）　　2.なし	1.あり　2.なし　3.不明
	その他	1.あり（内容　　処方量　　）　2.なし	1.あり　2.なし　3.不明
生物学的製剤	インフリキシマブ	1.あり（　　）mg/日　使用週（0,　,　,　週）（　　週毎） 2.なし	1.あり　2.なし　3.不明
	（複数回答可→）	[使用目的：1.皮膚病変　2.関節炎　3.その他（　　）]	
	その他	1.あり（薬剤名：　　）（用量:　　用法:　　） 2.なし	1.あり　2.なし　3.不明
	（複数回答可→）	[使用目的：1.皮膚病変　2.関節炎　3.その他（　　）]	
外用・光線療法等	副腎皮質ステロイド外用	1.あり　　2.なし	1.あり　2.なし　3.不明
	活性型ビタミンD3外用	1.あり　　2.なし	1.あり　2.なし　3.不明
	光線療法	1.あり[1.PUVA　2.NB-UVB　3.その他（　　）]　2.なし	1.あり　2.なし　3.不明
	その他	1.あり（内容　　量　　）　2.なし	1.あり　2.なし　3.不明

医療上の問題点

【WISH入力不要】

医療機関名

医療機関所在地

電話番号　　（　　）

医師の氏名

印　　記載年月日：平成　　年　　月　　日

20XX-XX-XX

28　表皮水疱症（接合部型及び栄養障害型）　臨床調査個人票（1. 新規）

ふりがな 氏名		性別	1. 男 2. 女	生年 月日	1. 明治 2. 大正 3. 昭和 4. 平成	年　月　日生 （満　歳）	
住所	郵便番号 電話　（　）			出生 都道府県		発病時在住 都道府県	
発病年月	1. 昭和 2. 平成　年　月（満　歳）	初診年月日	1. 昭和 2. 平成　年　月　日	保険種別	1. 政　2. 組　3. 船 4. 共　5. 国　6. 老		
身体障害者手帳	1. あり（等級＿＿＿級）　2. なし	介護認定	1. 要介護（要介護度＿＿＿）　2. 要支援　3. なし				
生活状況	社会活動（1. 就労　2. 就学　3. 家事労働　4. 在宅療養　5. 入院　6. 入所　7. その他（＿＿＿＿））						
	日常生活（1. 正常　2. やや不自由であるが独力で可能　3. 制限があり部分介助　4. 全面介助）						
家族歴	1. あり　2. なし　3. 不明 ありの場合（続柄　）	受診状況 （最近6か月）	1. 主に入院　2. 入院と通院半々　3. 主に通院（＿＿/月） 4. 往診あり　5. 入通院なし　6. その他（　）				

発症と経過（具体的に記述）

【WISH 入力不要】

疾患分類	1. 接合部型　（1. ヘルリッツ型　2. 非ヘルリッツ型　3. 幽門閉鎖合併型　4. その他） 2. 栄養障害型（1. 優性型　2. 劣性重症汎発型　3. その他） 3. その他　（　）
経過	1. ほぼ治癒　2. 軽快しつつある　3. 不変　4. 悪化しつつある　5. その他（　）

臨床所見（経過観察中に発現したものを含む）

皮膚粘膜症状

1. 水疱新生の数	1. 毎日　2. 週に4日以上　3. 週に3日以下　4. なし　5. 不明
2. 水疱・びらんの面積	1. 15%以上　2. 5%以上-15%未満　3. 5%未満　4. なし　5. 不明
3. 口腔内など粘膜の水疱・びらん	1. 毎日　2. 月に3、4日以上　3. 月に2日　4. なし　5. 不明
4. 水疱・びらん治癒後の瘢痕	1. あり　2. なし　3. 不明
5. 稗粒腫	1. あり　2. なし　3. 不明
6. 掌蹠角化	1. あり　2. なし　3. 不明
7. 脱毛	1. あり　2. なし　3. 不明
8. 爪変形・爪脱落	1. あり　2. なし　3. 不明

合併症

1. 貧血	1. あり（ヘモグロビン値：　g/dl）　2. なし　3. 不明
2. 低栄養	1. あり（血清アルブミン値：　g/dl）　2. なし　3. 不明
3. 高ガンマグロブリン血症	1. あり（血清IgG値：　mg/dl，IgA値：　mg/dl，IgM値：　mg/dl） 2. なし　3. 不明
4. 慢性炎症反応	1. あり（CRP値：　mg/dl）　2. なし　3. 不明
5. 全身性アミロイドーシス	1. あり（多臓器不全を伴う）2. あり（軽度臓器不全を伴う）　3. なし　4. 不明
6. 歯牙形成不全	1. あり　2. なし　3. 不明
7. 筋ジストロフィー	1. あり　2. なし　3. 不明
8. 肥厚性幽門狭窄	1. あり　2. なし　3. 不明
9. 指（趾）間癒着	1. 棍棒状癒着　2. 可動制限を伴う癒着　3. 可動制限の無い癒着 4. なし　5. 不明
10. 食道狭窄	1. 嚥下困難を伴う重度なもの　2. 軽度　3. なし　4. 不明 食道狭窄拡張術の施行（　回）
11. 扁平上皮癌の既往	1. あり（手術回数＿＿回）　2. なし　3. 不明
12. 腎機能障害	1. あり（血清クレアチニン　3mg/dl以上） 2. あり（血清クレアチニン　3mg/dl未満） 3. なし　4. 不明
13. 視力障害（矯正不能）	1. あり（両眼）　2. あり（片眼）　3. なし　4. 不明

病理学的事項					
水疱部	水疱初発位置	1. 表皮内	2. 接合部	3. 真皮内	
	基底細胞・有棘細胞の空胞変性	1. あり	2. なし	3. 不明	
非水疱部	係留線維の減少	1. あり	2. なし	3. 不明	
	半デスモソームの減少	1. あり	2. なし	3. 不明	
蛍光抗体法	ラミニン 332	1. 消失	2. 減弱	3. 普遍	4. 不明
	XVII 型コラーゲン	1. 消失	2. 減弱	3. 普遍	4. 不明
	α6β4インテグリン	1. 消失	2. 減弱	3. 普遍	4. 不明
	VII 型コラーゲン	1. 消失	2. 減弱	3. 普遍	4. 不明

遺伝子検査（実施している場合は記載してください。）

1. あり（患者：　　両親：　　）　2. なし

鑑別診断　下の疾患が鑑別できること

① 水疱性先天性魚鱗癬様紅皮症	1. 鑑別できる	2. 鑑別できない
② ポルフィリン症	1. 鑑別できる	2. 鑑別できない
③ 尋常性天疱瘡	1. 鑑別できる	2. 鑑別できない
④ 水疱性類天疱瘡	1. 鑑別できる	2. 鑑別できない
⑤ 線状 IgA 水疱性皮膚症	1. 鑑別できる	2. 鑑別できない
⑥ 疱疹状皮膚炎	1. 鑑別できる	2. 鑑別できない
⑦ 伝染性膿痂疹	1. 鑑別できる	2. 鑑別できない
⑧ 中毒性表皮壊死剥離症	1. 鑑別できる	2. 鑑別できない
⑨ 亜鉛欠乏による腸性肢端皮膚炎	1. 鑑別できる	2. 鑑別できない
⑩ 薬剤による水疱症	1. 鑑別できる	2. 鑑別できない
⑪ その他（　　）	1. 鑑別できる	2. 鑑別できない

医療上の問題点

【WISH入力不要】

医療機関名

医療機関所在地

医師の氏名　　電話番号　（　　）

印

記載年月日：平成　　年　　月　　日

（軽快者の症状が悪化した場合のみ記載）

症状が悪化したことを医師が確認した年月日　　平成　　年　　月　　日

特定疾患登録者証交付年月日　　平成　　年　　月　　日

20XX-XX-XX

28　表皮水疱症（接合部型及び栄養障害型）　臨床調査個人票（2. 更新）

ふりがな 氏　　名		性別	1. 男 2. 女	生　年 月　日	1. 明治 2. 大正 3. 昭和 4. 平成	年　月　日生 （満　歳）	
住　　所	郵便番号 電話　（　）			出　生 都道府県		発病時在住 都道府県	
発病年月	1. 昭和 2. 平成　年　月（満　歳）	初診年月日	1. 昭和 2. 平成　年　月　日	保険種別	1. 政　2. 組　3. 船 4. 共　5. 国　6. 老		
身体障害者手帳	1. あり（等級＿＿＿級）　2. なし	介護認定	1. 要介護（要介護度＿＿＿）　2. 要支援　3. なし				
生活状況	社会活動（1. 就労　2. 就学　3. 家事労働　4. 在宅療養　5. 入院　6. 入所　7. その他（＿＿＿）） 日常生活（1. 正常　2. やや不自由であるが独力で可能　3. 制限があり部分介助　4. 全面介助）					初回認定年月 1. 昭和 2. 平成　年　月	
受診状況（最近1年）	1. 主に入院　2. 入院と通院半々　3. 主に通院（＿＿/月）　4. 往診あり　5. 入通院なし　6. その他（　　）						

治療と経過（前回申請からの変化を中心に具体的に記述）

【WISH 入力不要】

疾患分類	1. 接合部型　（1. ヘルリッツ型　2. 非ヘルリッツ型　3. 幽門閉鎖合併型　4. その他） 2. 栄養障害型（1. 優性型　2. 劣性重症汎発型　3. その他） 3. その他　（　　）
経　過	1. ほぼ治癒　2. 軽快しつつある　3. 不変　4. 悪化しつつある　5. その他（　　）

臨床所見（経過観察中に発現したものを含む）

皮膚粘膜症状

1. 水疱新生の数	1. 毎日　2. 週に4日以上　3. 週に3日以下　4. なし　5. 不明
2. 水疱・びらんの面積	1. 15%以上　2. 5%以上-15%未満　3. 5%未満　4. なし　5. 不明
3. 口腔内など粘膜の水疱・びらん	1. 毎日　2. 月に3、4日以上　3. 月に2日　4. なし　5. 不明
4. 水疱・びらん治癒後の瘢痕	1. あり　2. なし　3. 不明
5. 稗粒腫	1. あり　2. なし　3. 不明
6. 掌蹠角化	1. あり　2. なし　3. 不明
7. 脱　毛	1. あり　2. なし　3. 不明
8. 爪変形・爪脱落	1. あり　2. なし　3. 不明

合併症

1. 貧血	1. あり（ヘモグロビン値：＿＿＿＿＿g/dl）　2. なし　3. 不明
2. 低栄養	1. あり（血清アルブミン値：＿＿＿＿＿g/dl）　2. なし　3. 不明
3. 高ガンマグロブリン血症	1. あり（血清IgG値：＿＿mg/dl，IgA値：＿＿mg/dl，IgM値：＿＿mg/dl） 2. なし　3. 不明
4. 慢性炎症反応	1. あり（CRP値：＿＿＿＿mg/dl）　2. なし　3. 不明
5. 全身性アミロイドーシス	1. あり（多臓器不全を伴う）2. あり（軽度臓器不全を伴う）3. なし　4. 不明
6. 歯牙形成不全	1. あり　2. なし　3. 不明
7. 筋ジストロフィー	1. あり　2. なし　3. 不明
8. 肥厚性幽門狭窄	1. あり　2. なし　3. 不明
9. 指（趾）間癒着	1. 棍棒状癒着　2. 可動制限を伴う癒着　3. 可動制限の無い癒着 4. なし　5. 不明
10. 食道狭窄	1. 嚥下困難を伴う重度なもの　2. 軽度　3. なし　4. 不明 食道狭窄拡張術の施行（　回）
11. 扁平上皮癌の既往	1. あり（手術回数＿＿＿回）　2. なし　3. 不明

12. 腎機能障害	1. あり（血清クレアチニン3mg/dl以上） 2. あり（血清クレアチニン3mg/dl未満）　3. なし　4. 不明
13. 視力障害（矯正不能）	1. あり（両眼）　2. あり（片眼）　3. なし　4. 不明

医療上の問題点

【WISH入力不要】

医療機関名

医療機関所在地

電話番号　（　）

医師の氏名

印　記載年月日：平成　年　月　日

20XX-XX-XX

［Ⅴ］

研究成果の刊行に関する一覧表

1．雑誌

欧文

著者名	論文題目	雑誌名	巻：頁，西暦年号
Hirai Y, Yamamoto T, Kimura H, Ito Y, Tsuji K, Miyake T, Morizane S, Suzuki D, Fujii K, Iwatsuki K	Hydroa vacciniforme is associated with increased numbers of Epstein-Barr virus-infected gdT-cells.	J Invest Dermatol	2012 Feb 7 [Epub ahead of print]
Fujii K, Aochi S, Takeshima C, Ohtsuka M, Hamada T, Asagoe K, Aoyama Y, Morizane S, Iwatsuki K	Eccrine Poromatosis Associated with Polychemotherapy.	Acta Dermatovenereol	2012 Feb 1 [Epub ahead of print]
Tanaka C, Hasegawa M, Fujimoto M, Iwatsuki K, Yamamoto T, Yamada K, Kawa K, Saikawa Y, Toga A, Mase S, Wada T, Takehara K, Yachie A	Phenotypic analysis in a case of hydroa vacciniforme-like eruptions associated with chronic active Epstein-Barr virus disease of gdT cells.	Br J Dermatol	166: 216-218, 2012
Fujii K, Suzuki N, Ikeda K, Hamada T, Yamamoto T, Kondo T, Iwatsuki K	Proteomic study identified HSP 70 kDa protein 1A as a possible therapeutic target, in combination with histone deacetylase inhibitors, for lymphoid neoplasms.	J Proteomics	75: 1401-1410, 2012
Yamasaki O, Morizane S, Aochi S, Ogawa K, Oono T, Iwatsuki K	Granulysin-producing cytotoxic T cells in the mucocutaneous lesions of Behcet disease: a distinct inflammatory response from erythema nodosum	Clin Exp Dermatol	36: 903-907, 2011
Ikeda K, Hamada T, Otsuka M, Iwatsuki K	Beneficial effects of neutrophil-targeted therapy for pyoderma gangrenosum associated with ulcerative colitis.	Eur J Dermatol	21: 804-805, 2011
Nakayama Y, Asagoe K, Yamauchi A, Yamamoto T, Shirafuji Y, Morizane S, Nakanishi G, Iwatsuki K	Dendritic cell subsets and immunological milieu in inflammatory human papilloma virus-related skin lesions.	J Dermatol Sci	63: 173-183, 2011
Matsuo T, Ichimura K, Tanaka T, Morizane S, Iwatsuki K, Eguchi M, Yoshino T	Bilateral conjunctival lesions in blastic plasmacytoid dendritic cell neoplasm.	Clin Exp Hematop	51: 49-55, 2011
Olsen EA, Whittaker S, Kim YH, Duvic M, Prince HM, Lessin SR, Wood GS, Willemze R, Demierre MF, Pimpinelli N, Bernengo MG, Ortiz-Romero PL, Bagot M, Estrach T, Guitart J, Knobler R, Sanches JA, Iwatsuki K, Sugaya M, Dummer R, Pittelkow M, Hoppe R, Parker S, Geskin L, Pinter-Brown L, Girardi M, Burg G, Ranki A, Vermeer M, Horwitz S, Heald P, Rosen S, Cerroni L, Dreno B, Vonderheid EC	Clinical Endpoints and Response Criteria in Mycosis Fungoides and Sézary Syndrome: a Consensus Statement of the International Society for Cutaneous Lymphomas (ISCL), the United States Cutaneous Lymphoma Consortium (USCLC) and the Cutaneous Lymphoma Task Force of the European Organization for Research and Treatment of Cancer (EORTC).	J Clin Oncol	29: 2598-2607, 2011
Kakimi K, Isobe M, Uenaka A, Wada H, Sato E, Doki Y, Nakajima J, Seto Y, Yamatsuji T, Naomoto Y, Shiraishi K, Takigawa N, Kiura K, Tsuji K, Iwatsuki K, Oka M, Pan L, Hoffman EW, Old LJ, Nakayama E	A phase I study of vaccination with NY-ESO-1f peptide mixed with Picibanil OK-432 and Montanide ISA-51 in patients with cancers expressing the NY-ESO-1 antigen.	Int J Cancer	129: 2836-2846, 2011

著者名	論文題目	雑誌名	巻：頁，西暦年号
Yamasaki O, Manabe K, Morimoto A, Iwatsuki K	Pustular erythema toxicum neonatorum in two siblings born to a mother with group B streptococcus colonization.	Eur J Dermatol	21: 271-272, 2011
Fujita A, Hamada T, Iwatsuki K	A retrospective analysis of 133 patients with cutaneous lymphomas from a single Japanese medical center between 1995 and 2008.	J Dermatol	38: 524-530, 2011
Nakai H, Sugata K, Usui C, Asano Y, Yamakita T, Matsunaga K, Mizokuchi Y, Katano H, Iwatsuki K, Yoshikawa T	A case of erythema multiforme associated with primary Epstein-Barr virus infection.	Pediatr Dermatol	28: 23-25, 2011
Takahashi H, Kouno M, Nagao K, Wada N, Hata T, Nishimoto S, Iwakura Y, Yoshimura A, Yamada T, Kuwana M, Fujii H, Koyasu S, Amagai M	Desmoglein 3-specific CD4+ T cells induce pemphigus vulgaris and interface dermatitis in mice	J Clin Invest	121: 3677-3688, 2011
Wada N, Nishifuji K, Yamada T, Kudoh J, Shimizu N, Matsumoto M, Peltonen L, Nagafuchi S, Amagai M	Aire-dependent thymic expression of desmoglein 3, the autoantigen in pemphigus vulgaris, and its role in T-cell tolerance	J Invest Dermatol	131: 410-417, 2011
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