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Blastic plasmacytoid dendritic cell neoplasm: a Swiss case series of a very rare disease and a structured review of the literature

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Summary

INTRODUCTION: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a very rare disease, with unique diagnostic challenges and often dismal outcome. There are no widely accepted treatment guidelines available. Lymphoma-like regimens with or without autologous or allogenic transplantation were the cornerstone of most therapeutic concepts. A few years ago, the CD123-directed immunoconjugate tagraxofusp emerged as a new valuable treatment option. The goal of our research was to collect available data on BPDCN-patients treated at large centres in Switzerland and worldwide and to draw conclusions regarding the incidence, clinical presentation, prognostic factors and therapeutic strategies.

METHODS: We collected data from BPDCN patients from leading Swiss haemato-oncology centres from 2005 to 2022. Furthermore, we reviewed and analysed the published literature (cohorts and case reports in peer-reviewed journals) from 1997 to 2020 (structured review of the literature).

RESULTS: We identified 115 international publications including 600 patients from all over the world. Most of them had very small sample sizes (only ten papers with more than ten patients) and all but one were retrospective or observational respectively. Most included patients were Europeans (n = 385, 64%) and Asians (n = 120, 20%), followed by Americans (n = 90, 15%) and patients from Australia/New Zealand (n = 3) and Africa (n = 2). BPDCN was more common in men with a predominance of 3:1. The median age (n = 414) at diagnosis was 66.5 years ranging from one month to 103 years. Newly diagnosed women were significantly younger than men (median: 62 vs 67 years, mean: 53.4 vs 59.3 years, p = 0.027) and less often had bone marrow infiltration and affected lymph nodes. Upfront allogenic transplantation as well as ALL regimens performed best, with response to first-line therapy clearly associated with better overall survival. The Swiss cohort contained 26 patients (23 males and 3 females) over 18 years (2005–2022). The median age at diagnosis was 68.5 years (range: 20–83). Ten patients underwent upfront stem cell transplantation (seven allogenic and three autologous), at least trending towards a better overall survival than other therapies. With a follow-up of 8 years, the median overall survival was 1.2 years. Eight patients in this cohort were treated with tagraxofusp, which became available in 2020 and was approved by Swissmedic in 2023.

CONCLUSIONS: Our study confirms that BPDCN is a very rare and difficult-to-treat disease. Underdiagnosis and underreporting in the literature pose further challenges. Symptoms at presentation seem to differ slightly between sexes and reaching a complete remission after first-line treatment remains crucial for a prolonged overall survival. Effective treatment protocols in first line include transplantation regimens (mainly allogenic, potentially also autologous) as well as ALL protocols. In order to understand the significance of tagraxofusp as a bridge to transplant or as a continuous monotherapy in elderly patients, further evaluation with longer follow-up periods is required. In general, analysis of the Swiss patients confirmed the results from the worldwide cohort.

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Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a very rare disease with an annual incidence of about 1 case

/ 2.5 million [1]. Patients usually present in their seventh decade; however all age groups are represented. A slight male predominance is well known with no ethnic predilection [2].

Uncertainties about the histogenesis of BPDCN have led to several changes in nomenclature in the past 25 years. At first, a T-cell origin was proposed based on the CD4 positivity of the cell. Hence BPDCN was named "plasmacytoid T-cell lymphoma". When the expression of CD56 and a lack of other T-cell markers on the cells were later documented, natural killer cells were regarded as the cells of origin. Accordingly, the WHO classification of 1999 classified the CD4+/CD56+ neoplasm as blastic natural killer cell lymphoma [3]. A few years later, Chaperot et al. [4] were able to demonstrate that these neoplastic cells expressing CD4/CD56 in the absence of typical markers of B-cell and T-cell lineage were derived from precursors of plasmacytoid dendritic cells. Based on these findings the term BPDCN was established and it has been used since 2008. The WHO classification of 2008 categorised BPDCN as an entity of "acute myeloid leukaemia and related precursor neoplasms". In the revised classification of 2016, BPDCN was finally classified as a separate entity [5]. The 5th and most recent edition of the WHO classification (2022) brought new changes, specifically revised diagnostic criteria. Currently, the diagnosis of BPDCN is established when one of the following two sets of criteria is met:

– expression of CD123 and one other pDC (plasmacytoid dendritic) marker (TCF4, TCL1, CD303, CD304) in addition to CD4 and/or CD56, or
– expression of any three pDC markers and absence of expression of all expected negative markers (CD3, CD14, CD19, CD34, lysozyme, myeloperoxidase) [6].

The clinical presentation is heterogeneous, but skin involvement at presentation is common, varying from nodular lesions to patch-plaques or even bruise-like patterns. In advanced stages of the disease, lymph node involvement, bone marrow infiltration and leukaemic transformations with marked cytopenia occur regularly [2].

The diagnosis is difficult and often missed or at least delayed. The cornerstone of the diagnosis is immunohistochemical work-up of skin biopsy or other affected tissues. BPDCN may be considered in cases of a morphologically diffuse, monomorphous infiltration of dermis with extension to subcutaneous fat (but not the epidermis however) of medium-sized blast cells with irregular nuclei and fine chromatin, some nucleoli, scant and agranular cytoplasm, variable mitosis, and the absence of angioinvasion and coagulative necrosis [5]. The diagnosis is confirmed by the expression of the typical markers, as mentioned above.

Nevertheless, differentiating neoplastic from reactive plasmacytoid dendritic cells is often very challenging, even for experienced haematologists. Several molecular and cytogenetic abnormalities have been reported, but none of them are unique to BPDCN. The diagnostic work-up/staging is completed by bone marrow examination and a PET/CT (currently not mandatory) or CT scan (with particular emphasis on lymph nodes and extramedullary disease [7]. Given the high incidence of occult cerebrospinal fluid involvement, a spinal fluid examination (including cytology and flow cytometry) is recommended in all patients [8].

Treatment of BPDCN is difficult and demanding, even though several treatment recommendations and position papers are now available [7–9].

In the past, most patients were treated with different chemotherapeutic regimens (acute lymphoblastic leukaemia-like, acute myeloid leukaemia-like, lymphoma-like). Despite high initial response rates, most patients relapse early and have dismal outcomes as reflected by the low median survival of only slightly above 12 months. This clinical course is usually independent of the initial pattern of the disease. Expression of CD303 and elevated Ki-67 might be associated with a longer survival [9]. Patients with a leukaemic stage have a worse prognosis with a median survival of 8.7 months [11]. Unequivocal curative concepts do not exist, even if long-term remissions in adults have been reported in selected cases with autologous or allogeneic stem cell transplantation [12]. Given that many patients are elderly, have additional comorbidities and present with advanced disease, therapeutic options are limited, especially regarding intensive approaches such as transplantation.

A promising new agent became available in 2019: tagraxofusp (Elzonris®, Stemline Therapeutics B.V.), a recombinant fusion protein consisting of human interleukin-3 fused to truncated diphtheria toxin that targets cells expressing CD123, leading to apoptosis [13]. Administration is by the intravenous route on days 1–5 of any 21-day cycle. Tagraxofusp is approved by the FDA, the EMA and Swissmedic and reimbursed by healthcare providers in Switzerland.

The goals of our study were to gain an overview of the prevalence, treatment and outcomes of Swiss patients with BPDCN in the last 18 years; to collect the available literature for the diagnosis and treatment of BPDCN; and to draw – in a cautious manner – conclusions about optimal management of this disease.

Materials and methods

We conducted extensive research of the available literature, mainly using the databases PubMed and Google Scholar as well as the relevant references in UpToDate. Our search terms were "BPDCN" and "blastic plasmacytoid dendritic cell neoplasm". Papers in English, German, French and Italian were considered. For every paper, we also checked the references to obtain further information about cohort studies or case reports involving patients with BPDCN. We obtained data published between 1 January 1997 and 31 December 2020. The key features of papers were entered in Excel spreadsheets, especially the following parameters: first author, type and geographic region of study, number of patients, age and sex distributions, clinical presentation at diagnosis, therapeutic regimen used, use of novel or experimental drugs, response rates, evolution and survival. Therapeutic regimens were divided into eight categories as follows: allogeneic transplantation (ALLO), autologous transplantation (AUTO), ALL-like without transplantation (ALL), AML-like without transplantation (AML), AL-like without further information without transplantation (AL NOS), lymphoma-like (LL), others (O), unknown (U). Table 1 lists the therapeutic

regimens included in each category, under consideration of sometimes blurred borders.

As we did not request the original raw data, our findings are based solely on the published information. The gathered information was analysed using descriptive as well as non-parametric statistics, the latter especially to compare outcomes of different therapeutic approaches. We quickly realised that investigating our pre-defined parameters was more difficult than expected, due to missing data and the inclusion of several patients in more than one report. In contrast to the descriptive analysis, exploratory statistics were only done in patient cases with availability of complete information as needed, maybe allowing a certain bias. The number of included patients is declared in a transparent manner on every separate analysis.

Statistical analyses were performed with Excel (Microsoft, Redmond, WA, USA), R 4.2.1 (www.r-project.org) and Vassarstats (Poughkeepsie, NY, USA). Categorical variables were compared between groups using Fisher's exact test. Continuous variables were compared between groups using Student's t-test for the international cohort and the Wilcoxon rank-sum test for the Swiss cohort. In the Swiss cohort, overall survival was calculated from diagnosis to death. Patients with no recorded death were censored at the date they were last known to be alive. In the structured review of the literature, we took the survival data from the original publications as far as possible. Overall survival was illustrated using the Kaplan-Meier methods and compared between groups using log-rank tests. The influence of covariables on overall survival was analysed using uni- and multivariate Cox regression models. For multivariate models, clinically relevant covariables were included as well as the therapy with the lowest hazard ratio, as due to collinearity only one therapy could be included.

In order to obtain an adequate overview of the Swiss BPDCN landscape, we contacted the leading local haemato-oncology centres asking for their data from BPDCN patients since 2005. The following centres (in alphabetical order) provided information: Cantonal Hospital Aarau, University Hospital Basel, Instituto Oncologico della Svizzera Italiana Bellinzona, University Hospital Inselspital Bern, Cantonal Hospital Chur, Cantonal Hospital Fribourg, University Hospital Geneva (HUG), University Hospital Lausanne (CHUV), Cantonal Hospital Baselland Liestal, Cantonal Hospital Lucerne, Cantonal Hospital St Gallen, Hirslanden Zurich, Municipal Hospital Zurich, University Hospital Zurich. For each patient, we collected data on the initial clinical presentation, histopathological features, the therapeutic regimen selected at first and in case of relapse, remission status and overall survival as well as epidemiological data (sex, age at diagnosis). Of note, the same eight therapeutic categories were maintained as in the international structured review of the literature. Transplant (ALLO or AUTO) as first-line treatment means: The decision to do an allogenic (autologous) stem cell transplantation was taken before starting treatment with induction therapy

Table 1:
Categories of therapeutic regimens.

Allogeneic stem cell transplantation (ALLO)	Induction therapies containing cytarabine/anthracycline ± intrathecal cytarabine, CHOP, MTX, etoposide, cytarabine
Autologous stem cell transplantation (AUTO)	Induction therapy with CHOP
Acute lymphatic leukaemia (ALL)	Ifosfamid, etoposide, prednisone ± cytarabine ± MTX ± L-asparaginase ± mitoxantrone
	Hyper-CVAD
	VPDL (vincristine, methylprednisolone, daunorubicin, L-asparaginase) ± MTX
	LALA 94 protocol
	GRAALL 2003 protocol
Acute myeloid leukaemia (AML)	Cytarabine/anthracycline ± intrathecal MTX ± etoposide ± mitoxantrone ± fludarabine ± lomustine ± vincristine
	LAM-90 protocol
Acute leukaemia not otherwise specified (AL NOS)	No specified protocols
Lymphoma-like (LL)	CHOP-like (COP ± anthracycline ± rituximab ± etoposide ± bleomycin ± chlorambucil ± MTX ± mitoxantrone ± intrathecal MTX)
	Hyper-CVAD
	ABVD
	IE, ICE or IME (MTX instead of carboplatin)
	ESHAP
Other therapeutic options (O) in alphabetical order	Azacytidine
	Bendamustine
	Best supportive care / no treatment
	Cytarabine ± mitoxantrone ± etoposide
	Daratumumab
	EC (epirubicin, cyclophosphamide)
	Hydroxyurea
	Interferon alpha ± bexarotene
	POMP-like
	Radiotherapy ± etoposide
	Steroid monotherapy
	Tagraxofusp
	Unspecified chemotherapy
	VAD (vincristine, anthracycline, dexamethasone) plus radiotherapy
	Venetoclax
	VRd (bortezomib, lenalidomide, dexamethasone)

(and not at the time point when 1st line therapy failed, then ALLO or AUTO were counted as 2nd line treatment). All data were provided in an anonymised form and the study was approved by the local ethics committee of Nordwest- und Zentralschweiz EKNZ (Nr. 2016/271). A study protocol was not prepared. All study data are available to those with a legitimate interest (Open Science).

Results

Swiss cohort

We obtained data from 26 patients diagnosed with blastic plasmacytoid dendritic cell neoplasm (BPDCN) in the Swiss centres since 2005. Another two patients (both female) had to be excluded from analysis due to missing/denied general consent (table 2).

Two examples of patient cases are described in figure 1. The median age of patients was 68.5 years (range: 20–83). Out of these patients, 23 (89%) were male and 3 female.

Skin manifestations at diagnosis were observed in 21 patients (81%), lymphadenopathy in 12 patients (46%), bone marrow involvement in 17 patients (65%), and blasts were detected in the peripheral blood in 11 cases (42%). Given the substantial male predominance of our cohort, no conclusions regarding sex-based differences could be drawn.

Figure 1: Examples of two Swiss patients. (**A**) Patient #11: this 72-year-old man presented with affection of skin, lymph nodes, bone marrow, peripheral blood and spleen. He underwent treatment with autologous stem cell transplantation according to GRAALL-2012 protocol and achieved complete remission. (**B**) Patient #14: This 71-year-old man initially presented with a 6-month history of skin lesions. Based on the clinical and laboratory findings, the patient was diagnosed with blastic plasmacytoid dendritic cell neoplasm with only skin manifestations. Due to age and co-morbidities, allogeneic stem cell transplantation was no option. Therefore, a "wait and see" approach was adopted. Only one month later transformation into acute leukaemia was observed. Chemotherapy with B-CHOP (bortezomibe, cyclophosphamide, vincristine, adriablastine, prednisolone) was initiated, resulting in a very short partial response and early disease progression, leading to introduction of a 2nd line therapy with ABVD. Unfortunately, the patient passed away due to complications from an intracerebral bleeding, 9 months after the diagnosis and 15 months after the onset of the first clinical symptoms.

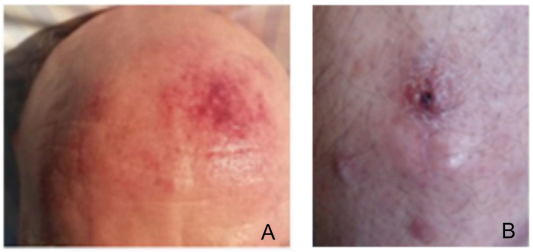


Table 2:
Overview of the Swiss cohort with 26 patients.

Patient #	Sex / age (years)	Clinical picture at diagnosis				Therapeutic regimen after diagnosis	First-line therapy → Response (→ Relapse)	Overall survival (months)
		Skin	Lymph node	Bone marrow	Peripheral blood			
1	M/53	yes	no	yes	no	ALLO-SCT (AML protocol)	ALLO → CR	100.9*
2	M/61	yes	no	no	no	ALLO-SCT (AML protocol)	ALLO → CR	75.5**
3	M/20	yes	no	no	no	CODOX-M IVAC (LL protocol)	LL → CR → Relapse	6.2
4	M/78	yes	yes	yes	yes	Pralatrexate (LL)	LL → PD	8.1
5	M/40	no	no	yes	no	ALLO-SCT (cytarabine, idarubicin)	ALLO → CR → Relapse	30
6	F/75	no	no	yes	yes	Azacytidine (AML protocol)	AML → PD	7.1
7	M/68	no	no	yes	no	GMALL elderly (ALL protocol)	ALL → PD	3.9
8	M/74	yes	no	no	no	CHOEP (LL)	LL → PR → PD	10.8
9	M/44	no	yes	no	no	ALLO-SCT (hyper-CVAD, HD-MTX/cytarabine)	ALLO → CR	97.8*
10	M/81	yes	yes	yes	yes	3 cycles tagraxofusp	O*** → CR → Relapse	10.3**
11	M/72	yes	yes	yes	yes	AUTO-SCT (GRAALL-2014)	AUTO → CR	45.6*
12	M/47	yes	yes	yes	yes	1 cycle hd cytarabine, 2 cycles tagraxofusp (complete remission after 1 cycle)	O*** → CR → Relapse	1.9
13	F/69	yes	yes	yes	no	2 cycles cytarabine/daunorubicin (AML protocol)	AML → PD	4.6
14	M/71	yes	no	no	no	None (only skin manifestation)	O → PD	9.2
15	M/73	yes	no	n.d.	no	3 cycles tagraxofusp	O*** → CR	17.7*
16	M/68	yes	no	no	no	AUTO-SCT (hyper-CVAD)	AUTO → CR	87.3
17	M/83	yes	no	no	no	None (only skin manifestation)	O → PD	56.5
18	F/67	yes	no	no	no	AUTO-SCT (hyper-CVAD)	AUTO → CR	110.6**
19	M/56	yes	yes	yes	yes	ALLO (GRAALL-2014B)	ALLO → CR	79.6*
20	M/53	no	yes	yes	yes	ALLO-SCT (Hyper-CVAD)	ALLO → CR → Relapse	4.5
21	M/70	yes	yes	yes	yes	5 cycles tagraxofusp	O*** → PR → Progress	11.9
22	M/21	yes	yes	yes	yes	1 cycle tagraxofusp	O*** → PD → ALLO	33.1*
23	M/69	yes	yes	yes	no	Prednisone/cyclophosphamide, 2 cycles Tagraxofusp	O*** → CR → Relapse	9.6
24	M/53	yes	no	yes	no	ALLO (3 cycles tagraxofusp)	ALLO → CR → Relapse	17.7*
25	M/82	yes	yes	yes	yes	AML (venetoclax, azacytidine)	AML → PD	13.9
26	M/78	yes	no	yes	yes	5 cycles tagraxofusp	O*** → CR → Relapse	15.8

ALL: acute lymphatic leukaemia-like without transplantation; ALLO: allogeneic transplantation; AML: acute myeloid leukaemia-like without transplantation; AUTO: autologous transplantation; CR: complete remission; LL: lymphoma-like; O: other therapeutic options; PD: progressive disease; PR: partial remission; SCT: stem cell transplantation.

* Alive at last contact.

** Death due to reason other than blastic plasmacytoid dendritic cell neoplasm.

*** tagraxofusp.

Ten patients (38%) received a stem cell transplantation as first-line therapy (seven ALLO and three AUTO). Of the seven patients who underwent ALLO, the induction regimens were as follows: 3 AML-like, 1 ALL-like, 2 lymphoma-like and 1 Other (tagraxofusp).

The remaining 16 patients received 1st line treatment as follows: 1 ALL, 3 AML, 3 LL, 9 Other (7 tagraxofusp, 2 Watch -and-Wait).

In brief, the responses achieved by the seven patients with tagraxofusp plus one patient with tagraxofusp as induction therapy before ALLO were as follows:

Patient #10 achieved a complete remission after only 3 cycles without further therapy. He relapsed approximately 8 months after diagnosis and died due to COVID-19 shortly after.

Patient #12 received an induction with 1 cycle of high-dose cytarabine, followed by 2 cycles of tagraxofusp. Despite achieving a complete remission after the 1st cycle of tagraxofusp, he then progressed and died without further therapy.

Patient #15 received 3 cycles of tagraxofusp – complicated by severe capillary leak syndrome – and achieved a complete remission. Without further treatment he was alive with overall survival 17.7 months at last contact.

Patient #21 achieved a PR after 5 cycles of tagraxofusp. As treatment had to be stopped for healthcare-insurance issues, he rapidly progressed and died without further therapy.

Patient #22 progressed after 1 cycle of tagraxofusp, was stabilised with 2 cycles of Hyper-CVAD and then underwent an ALLO. He was still alive 33 months after diagnosis.

Patient #23 received 2 cycles of tagraxofusp after short induction therapy with cyclophosphamide and prednisone, achieving a complete remission after one cycle. Due to progression after the 2nd cycle, salvage therapy with azacytidine and venetoclax was started. The patient died after the 4th cycle.

Patient #24 received 3 cycles of tagraxofusp as induction before ALLO which resulted in a complete remission. Seven months later, he relapsed and was treated with Hyper-CVAD before emigrating to his homeland. He was still alive 6 months after starting salvage therapy.

Patient #26 achieved a complete remission after 3 cycles of tagraxofusp. After 5 cycles, he progressed and was treated with seven cycles of azacytidine and venetoclax, dying thereafter due to refractory disease.

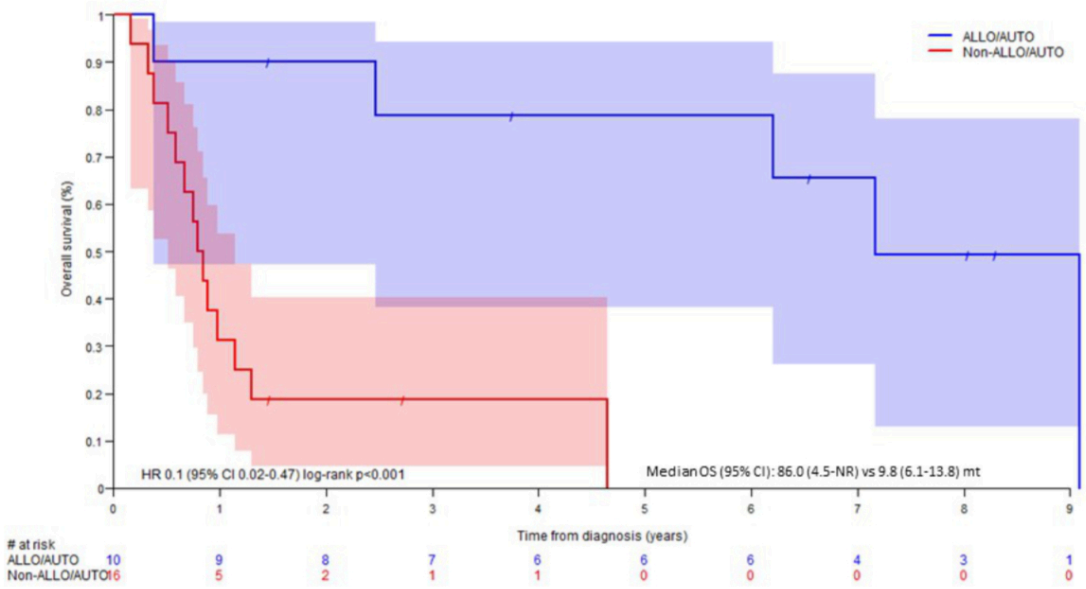
Two other cases deserve special attention, as they presented with asymptomatic skin lesions only, leading to an initial Watch-and-Wait approach. Patient #17 was diagnosed in 2011 at the age of 83 years. Four years later, he transformed to AML, salvage therapy with hydroxyurea and later decitabine was initiated without effect and the patient passed away. Patient #14 is described in figure 1.

All seven patients who underwent ALLO as first-line along with the three patients who underwent AUTO achieved complete remission, compared to only 38% (n = 6) of the 16 patients without transplantation (p = 0.009). Considering overall survival, 1st line therapy with AUTO/ALLO had much better outcomes than the rest (HR: 0.1, 95% CI: 0.02–0.47, p <0.001, figure 2) with a median overall survival of 86 months (4.5–NR) vs 9.8 (6.1–13.8) months.

The seven patients still alive were treated with ALLO (four patients, one of them with tagraxofusp – induction), tagraxofusp (two patients, 1 of them progressed and underwent ALLO as 2nd line) and AUTO (one patient).

The median follow-up time was 8 years. The median overall survival was 1.2 years (95% CI: 0.8–6.2 years) (figure 3). The multivariate analysis revealed a significant association between survival and involvement of skin (HR: 0.22, 95% CI: 0.06–0.82, p = 0.024) and 1st line therapy (ALLO/AUTO vs non-ALLO/AUTO, HR: 0.06, 95% CI: 0.01–0.32, p = 0.001). Patients with skin involvement at

Figure 2: Kaplan-Meier curve of overall survival in the Swiss cohort according to therapy.



diagnosis and treatment with ALLO/AUTO achieved a particularly good outcome (small numbers, data not shown).

Structured review of the literature

We identified 115 international studies (listed in the appendix) including 600 patients from 1997 to 2020.

Most of the studies were case reports with fewer than ten patients. Details regarding therapeutic protocols and outcomes are lacking in many studies with larger cohorts [11, 14–22].

Most included patients were Europeans (n = 385, 64%) and Asians (n = 120, 20%), followed by Americans (n = 90, 15%) and patients from Australia/New Zealand (n = 3) and Africa (n = 2). Of the 115 publications, 96 (148 patients) were case reports and 18 (405 patients) retrospective cohort analyses. We found only one (47 patients) prospective study (the first phase 1/2 trial with tagraxofusp [13]).

BPDCN was more common in men with a predominance of 3:1. The median age (n = 414) at diagnosis was 66.5 years (ranging from 1 month to 103 years). Of note, women were significantly younger than men (median: 62 vs 67 years, mean: 53.4 vs 59.3 years, p = 0.027).

The clinical picture at diagnosis was heterogeneous (n = 516): skin lesions were observed in 249 patients (missing data in 219 patients), lymphadenopathy in 94 (missing data in 309 patients), bone marrow infiltration in 116 (missing data in 309 patients) and blasts in peripheral blood as an expression of leukaemic state in 47 (missing data in 309 patients). Based on sex, there were significant differences in the clinical presentation regarding bone marrow infiltration (p = 0.005) and lymph node involvement (p = 0.022) with both being more common in males (table 3).

Other manifestations affecting <10% of patients included hepatomegaly (7.8%), splenomegaly (9.1%), central nervous system involvement (5.4%), or lung and pleural involvement (1.9%). However, almost any organ could be involved in individual cases (such as chest wall, gastrointestinal tract, conjunctiva, testis, buttock, ovaries, paravertebral tissue, parotid gland, lacrimal gland and thyroid gland, gallbladder, orbital cavity).

Unfortunately, we found no / very little data on the time elapsed between the onset of the first symptoms and the diagnosis.

We tried to assign each of the 516 patients to a therapeutic regimen (figure 4). ALLO was more prevalent in Europe (18% of European patients), Asia and America. Most patients in America were treated with ALL, while AML was predominantly performed in Europe. LL were in use in all continents in about 15–25% of patients. As (unlike in the Swiss Cohort) the primary attribution could not be undertaken with sufficient certainty, only 357 patients were considered for demonstrating the distribution of different 1st

Figure 3: Kaplan-Meier curve of overall survival in the Swiss cohort.

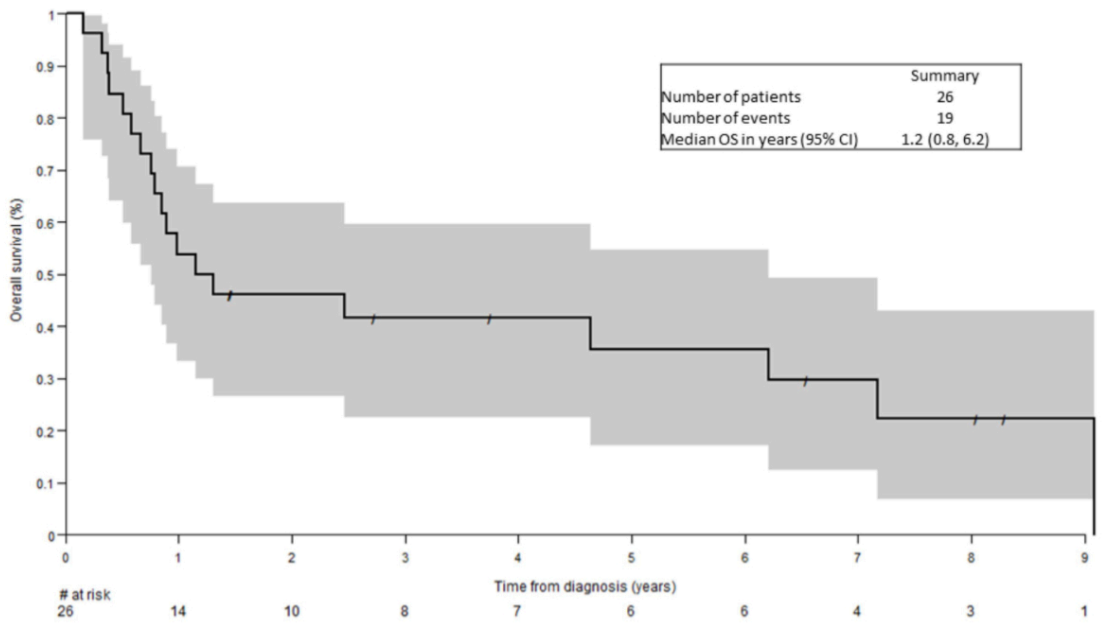


Table 3:
Differences in presentation by sex.

	Females (n = 119)	Males (n = 295)	Missing data	Fisher's exact test
Involved organ	**n (%)**	**n (%)**	**n**	**p-value (2-tailed)**
Bone marrow	25/57 (43.9%)	91/148 (61.5%)	209	0.028
Lymph nodes	19/59 (32.2%)	75/147 (51.0%)	208	0.02
Peripheral blood (blasts)	9/58 (15.5%)	38/147 (25.9%)	209	0.141
Skin	71/87 (81.6%)	178/210 (84.8%)	117	0.493

line therapeutic regimens. The most used regimens were ALLO (87 or 24%), followed by LL (83 or 23%), ALL (49 or 14%), AML (28 or 8%), AUTO (15 or 4%) and AL NOS (11 or 3%). A quarter of patients (84 or 24%) received a different treatment ("Other" category). Survival data were available for 364 patients. The median overall survival was 15 months (95% CI: 13–18, figure 5A) with no significant differences between sexes (p = 0.13, figure 5B).

Achieving a complete remission was significantly associated with a longer overall survival (figure 5C). Upfront transplant-concepts resulted in a high complete remission rate, e.g. ALLO vs non-ALLO (71.3% vs 57.8%, p <0.001) and ALLO/AUTO vs non-ALLO/AUTO (78.6% vs 26.0%, p = 0.001), ALL vs non-ALL (81.6% vs 57.8%, p = 0.026). AUTO vs non-AUTO was not meaningful due to small sample size. Overall, more intensive approaches like ALLO or ALL were associated with a longer overall survival (figure 5D; only therapeutic regimens with sufficient sample sizes are considered). ALL was non-inferior to ALLO in terms of overall survival (p = 0.416).

In a univariate analysis, overall survival was determined by age, involvement of bone marrow and other organs, blasts in the peripheral blood, choice of 1st line therapy and response to 1st line therapy (table 4).

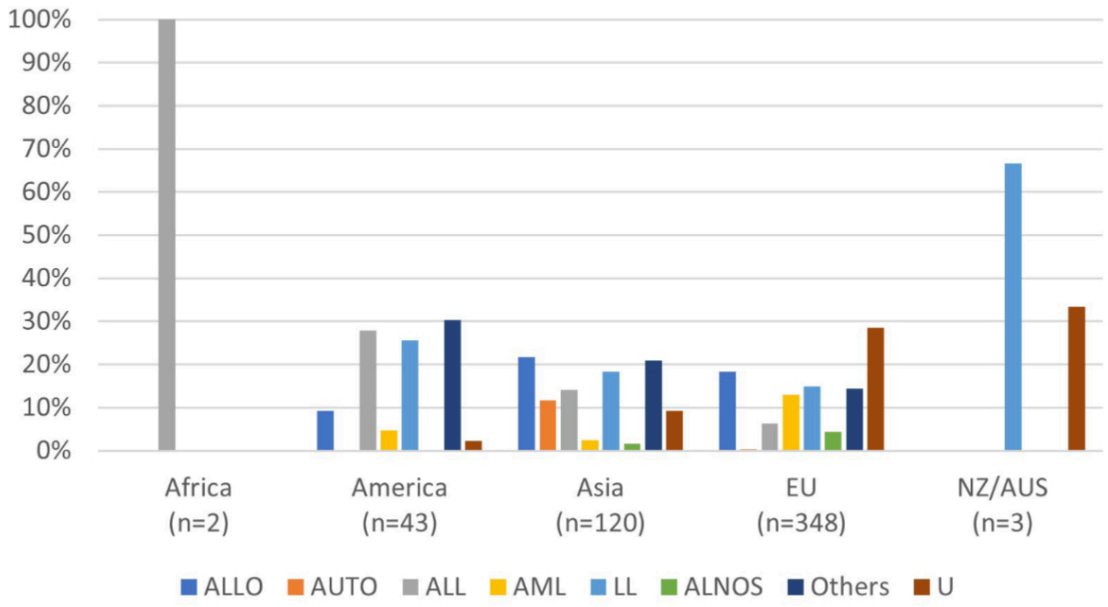
The multivariate analysis showed that in addition to the treatment concept, achieving a complete remission in first line was the strongest additional prognostic factor, and that bone marrow infiltration at diagnosis had a negative impact on overall survival (table 5).

Discussion

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a very rare disease with an often dismal outcome due to prolonged time to diagnosis, and lack of effective therapies and of randomised controlled prospective trials. We expect around 3-4 new diagnoses in Switzerland every year. The available literature mainly consists of case reports, case series and retrospective analyses. Phase 3 trials are lacking because of the rarity of the disease and the absence of recognised therapeutic options. The clinical presentation is heterogeneous, with cutaneous lesions often serving as a key to diagnosis, followed by lymphadenopathies, bone marrow involvement, cytopenia and leukaemic transformation.

Our cohort of Swiss patients contained 26 patients (two additional patients were not included in the analyses due to lack of informed consent) in 18 years with a median fol-

Figure 4: Geographic distribution of therapeutic regimens. U: unknown.



	AFRICA		AMERICA		ASIA		EUROPE		AUSTRALIA			Total	
ALLO			4	9%	26	22%	64	18%				94	18%
AUTO					14	12%	1	0%				15	3%
ALL	2	100%	12	28%	17	14%	22	6%				53	10%
AML			2	5%	3	3%	45	13%				50	10%
LL			11	26%	22	18%	52	15%	2	67%		87	17%
AL NOS					2	2%	15	4%		0%		17	3%
Others			13	30%	25	21%	50	14%		0%		88	17%
U			1	2%	11	9%	99	28%	1	33%		112	22%
Total	2		43		120		348		3			516	100%

low-up of 8 years. The mean population in Switzerland during this period was 8.1 million, yielding an incidence of 0.19 / million / year, which is slightly less than expected. The median age of onset, the male predominance, clinical presentations and survival curves are in line with previous findings as demonstrated in our structured literature review (see below).

Figure 5: Overall survival according to different conditions. (A) Overall survival – structured review of the literature; (B) Overall survival according to sex; (C) Overall survival according to remission status; (D) Overall survival according to therapeutic regimens.

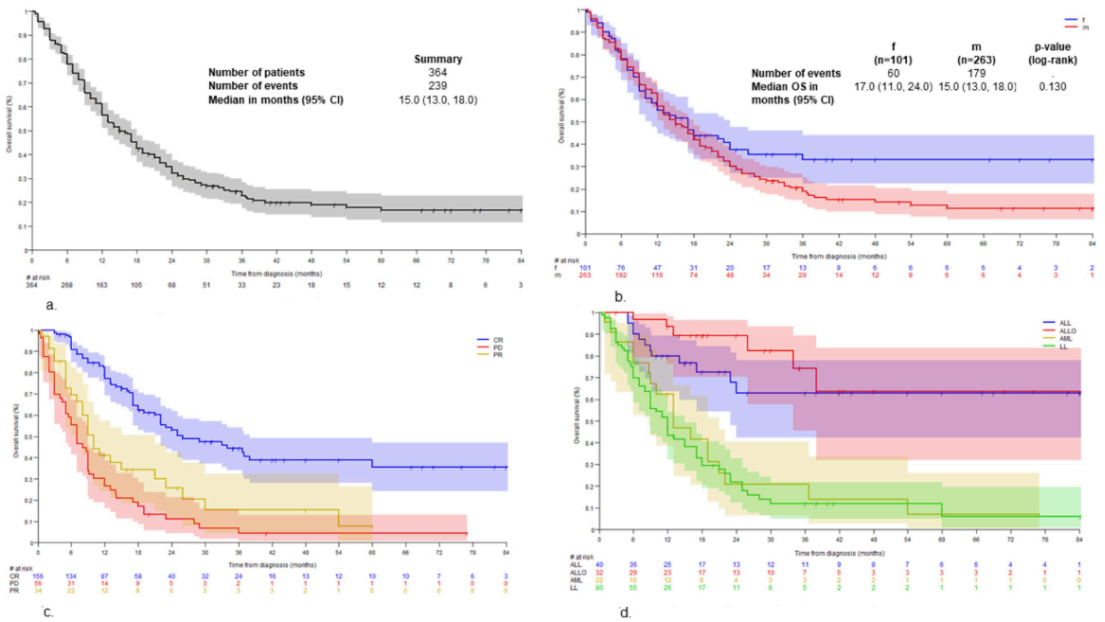


Table 4:
Univariate analysis of parameters influencing overall survival.

Variable	n	HR (95% CI)	p-value
Age (years)	414	1.03 (1.02–1.04)	<0.001
Sex (M vs F)	414	1.25 (0.93–1.68)	0.134
Skin (Yes vs No)	297	1.02 (0.67–1.56)	0.922
Lymph nodes (Yes vs No)	206	1.45 (0.97–2.15)	0.068
Bone marrow (Yes vs No)	205	1.53 (1.02–2.31)	0.042
Peripheral blood (Yes vs No)	205	1.72 (1.11–2.67)	0.016
Other organ involvement (Yes vs No)	206	1.66 (1.07–2.56)	0.023
Complete remission (Yes vs No)	349	0.33 (0.25–0.44)	<0.001
Relapse (Yes vs No)	193	1.45 (0.98–2.14)	0.060
Leukaemic transformation (Yes vs No)	155	1.74 (0.99–3.05)	0.054
Variable (1 st line treatment)			
ALLO/AUTO vs non-ALLO/AUTO	254	0.18 (0.09–0.39)	<0.001
ALLO/AUTO/ALL vs non-ALLO/AUTO/ALL	254	0.19 (0.11–0.30)	<0.001
ALLO vs non-ALLO	254	0.21 (0.10–0.45)	<0.001
ALL/AUTO vs non-ALL/AUTO	254	0.27 (0.15–0.49)	<0.001
ALL vs non-ALL	254	0.31 (0.17–0.55)	<0.001

ALL: acute lymphatic leukaemia; ALLO: allogeneic transplantation; AUTO: autologous transplantation.

Table 5:
Multivariate analysis of parameters influencing overall survival.

Variable	HR (95% CI)	p-value
Complete remission (yes vs no)	0.06 (0.02–0.18)	<0.001
ALLO/AUTO vs non-ALLO/AUTO	0.09 (0.02–0.38)	<0.001
Relapse (yes vs no)	8.63 (2.89–25.76)	<0.001
Bone marrow (yes vs no)	2.24 (1.28–3.92)	0.005
Age (years)	1.01 (1.00–1.02)	0.094
Lymph nodes (yes vs no)	1.44 (0.86–2.40)	0.163
Peripheral blood (yes vs no)	1.20 (0.60–2.39)	0.606
Leukaemic transformation (yes vs no)	1.19 (0.56–2.53)	0.658

ALLO: allogeneic transplantation; AUTO: autologous transplantation.

The rate of patients undergoing allogeneic transplantation is similar (Switzerland: 31%, structured review of the literature: 24%), as is the general heterogeneity of therapeutic regimens in use. Due to the small sample size, our findings were not statistically significant. However, we made some interesting observations. All seven patients with ALLO as first line achieved a complete remission; the same holds true for the 3 AUTO patients. Only one ALL-regimen upfront was chosen in Switzerland (PD). Of 3 LL, only one led to a complete remission (1 PR, 1 PD) and all three AML (without consolidating ALLO) resulted in PD. The nine other first-line strategies mainly contained tagraxofusp (seven patients; 5 complete remission, 1 PR, 1 PD) or Watch-and-Wait (because of modest skin manifestation only, n = 2). Nearly all patients (with two exceptions) diagnosed in 2020 or later had tagraxofusp as part of their treatment. The follow-up is still too short to comment on the efficacy of tagraxofusp. The treatment regimen in the relapsed setting contained hydroxyurea, ALL protocols, B-CHOP, ABVD, hypomethylating agents and venetoclax. Of the 26 patients, seven are still alive (4 ALLO, 1 AUTO, 1 tagraxofusp) including one patient with ALLO 2nd line after progression to tagraxofusp and salvage treatment with Hyper-CVAD.

The structured review of the literature confirmed that BPDCN is mainly a disease of older age, even if it can occur at any age. Male patients seemed to more often have bone marrow infiltration and lymphadenopathies at diagnosis than women, a fact not affecting the outcome in a statistically significant manner. Median overall survival was around 15 months. Assuming underdiagnosing and underreporting of BPDCN, overall survival might even be shorter. Subdividing the therapeutic approaches into six different groups, upfront allogenic concepts or ALL regimens clearly performed better than AML-like or lymphoma-like treatments. The same probably holds true for an upfront autologous concept, even if the numbers were too small to draw firm conclusions. For the latter reason, the group with Other treatments could not be subdivided and no serious evaluation of efficacy was possible. Our study confirms that deeper responses to first-line therapy are cornerstones of a better outcome with patients in complete remission achieving the longest overall survival.

Different new therapeutic strategies involving targeted therapies have been increasingly used over the last decade. Bortezomib, an inhibitor of the nuclear factor-kappa B pathway, demonstrated promising results in combination with chemotherapy in mice [23]. The combination of lenalidomide and dexamethasone was successfully used in several case reports [24]. In a case at our institution, the use of bortezomib in combination with CHOP did not result in long-term remission.

The anti-CD38 antibody daratumumab was also used with mixed results, since CD38 is not only expressed on lymphoid tissues (mainly plasma cells), but also on myeloid cells [25]. Immunohistochemical investigations of BPDCN biopsies revealed [25] prominent staining of the anti-apoptotic protein BCL-2 leading to trials with venetoclax, with no measurable success. The discovery of PD-L1 positivity in approximately 50% of investigated BPDCN samples (range: 1% to 55%) has raised hopes about the potential of checkpoint inhibitors against PD-L1, although no such reports have been published to date [27]. Single reports of anti-CD123-CAR T-cell therapy have been published with discordant outcomes [28].

Tagraxofusp entered the scene a few years ago as the “new kid on the block”. This CD123-directed cytotoxin led to a high ORR, serving as a perfect induction – or bridging – therapy to transplant in eligible patients. A relevant and potentially lethal side effect is capillary leak syndrome. Nevertheless, tagraxofusp has become a new standard of care for patients with BPDCN and was approved by the Swiss authorities (Swissmedic) by January 2024. As of today, allogenic stem cell transplantation seems to be the best therapeutic option in patients with BPDCN, especially in younger patients with a good performance status. Complete remission can be achieved in about 60% of these patients. Future directions will probably focus on the development of more appropriate (preferably allogenic) CAR T-cell therapy or bispecific antibodies, as well as on combination therapies including tagraxofusp.

The strength of our study is the extensive work-up of available literature, and this not only in the English language, but also in German, French and Italian. It gives a comprehensive overview of BPDCN as a rare disease entity in the national and worldwide context. By including the largest haemato-oncology centres in Switzerland, a substantiated statement about prevalence and therapeutic modalities in Switzerland can be given.

Our study has several limitations. First, the data drawn from the 115 international papers were often not complete, not related to the scope of the present study and of heterogeneous quality. Since, as mentioned, the term BPDCN was only introduced in 2008, it cannot be ruled out that we would have found further (especially older) relevant literature with extended search terms. As such, stringent conclusions beyond a descriptional and hypothesis-generating manner cannot be drawn from this kind of even structured literature review. Secondly, due to upcoming availability of tagraxofusp, original articles and case reports as well as cohort studies were only considered up to 2020. Thirdly, collecting the data for our Swiss cohort in a retrospective manner meant that not every centre could provide us with required details, for example the time from first symptoms to diagnosis and treatment. Further, it cannot be excluded that a few BPDCN patients were treated outside the larger centres, even if the probability is low. As the number of patients in the Swiss cohort is very small, it is difficult to demonstrate statistically significant differences between subgroups.

In conclusion, our study yielded deeper insights into a rare, heterogeneous disease, which is difficult to treat and where comparative studies are lacking. We propose that national or even continental registries be established for every orphan disease.

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Potential competing interests

All authors have completed and submitted the International Committee of Medical Journal Editors form for disclosure of potential conflicts of interest. No potential conflict of interest related to the content of this manuscript was disclosed.

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Appendix

Structured Review of the literature_Publications

1. **Roodbergen, SL, J Hofland, KH Lam, PK Dikrama, A Broyl, and K Monkhorst**, *Blastic plasmacytoid dendritic cell neoplasm.* Br J Haematol, 2014. 164(6): p. 757.
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