

Supplementary material

Search Strategy

We conducted a comprehensive search across PubMed, EMBASE, Cochrane Central Register of Controlled Trials, and Web of Science Core Collection on 14th December 2023.

PubMed

(inflamm*[Title]) OR (Behcet's[Title] OR Behçet's[Title] OR behcet[Title] OR Behçet[Title] OR Behcet Syndrome[Title]) OR (myelodysplastic syndromes[Title] OR myelodysplastic syndrome[Title] OR MDS[Title]) AND (Trisomy 8).
(n=288).

EMBASE

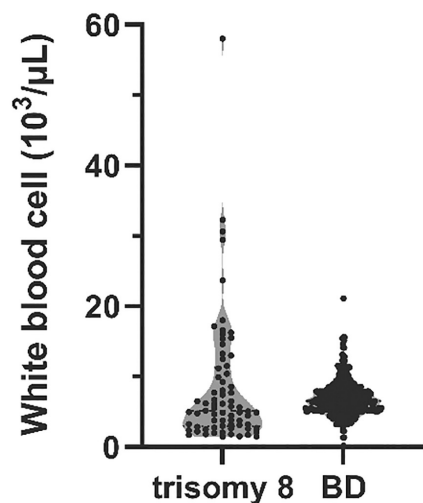
ti((inflamm*) OR (Behcet's OR Behçet's OR behcet OR Behçet OR Behcet Syndrome) OR (myelodysplastic syndromes OR myelodysplastic syndrome OR MDS)) AND (Trisomy 8).
(n=906).

Cochrane Central Register of Controlled Trials

#1 (inflamm*):ti OR (Behcet's OR Behçet's OR behcet OR Behçet OR Behcet Syndrome):ti OR (myelodysplastic syndromes OR myelodysplastic syndrome OR MDS):ti
#2 (Trisomy 8).
#3 (#1 AND #2).
(n=16).

Web of Science Core Collection

TI=((inflamm*) OR (Behcet's OR Behçet's OR behcet OR Behçet OR Behcet Syndrome) OR (myelodysplastic syndromes OR myelodysplastic syndrome OR MDS)) AND (Trisomy 8).
(n=332).



Supplementary Fig. S1. Distribution of white blood cell counts.

Trisomy 8 (n=73): Mean ± SD: 8.57±9.06, Median: 5.17.

BD (n=420): Mean ± SD: 6.93±2.40, Median: 6.60.

Gene		Patients														
		1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
Myeloid Sequencing Panel	ABL1															
	ASXL1															
	ATRX															
	BCOR															
	BCORL1															
	BRAF															
	CALR															
	CBL															
	CBLB															
	CBLG															
	CDKN2A															
	CEBPA															
	CSF3R															
	CUX1															
	DNMT3A															
	ETV6/TEL															
	EZH2															
	FBXW7															
	FLT3															
	GATA1															
	GATA2															
	GNAS															
	HRAS															
	IDH1															
	IDH2															
	IKZF1															
	JAK2															
	JAK3															
	KDM6A															
	KIT															
	KRAS															
	MLL															
	MPL															
	MYD88															
	NOTCH1															
	NPM1															
	NRAS															
	PDGFRA															
	PHF6															
	PTEN															
	PTPN11															
	RAD21															
	RUNX1															
	SETBP1															
	SF3B1															
	SMC1A															
	SMC3															
	SRSF2															
	STAG2															
	TET2															
	TP53															
	U2AF1															
	WT1															
	ZRSR2															
MEFV gene	M694I															
	M680I															
	M694V															
	V726A															
	E84K															
	E148Q															
	L110P-E148Q															
Inflammasome	P369S-R408Q															
	R202Q															
	G304R															
	S503C															
	NALP3															

Supplemental Fig. S2. Mutation genes identified in cases in the systematic review.

Gene mutations were indicated in red (myeloid disorder), blue (MEFV), and green (inflammasome).

Supplementary Table S1. Baseline characteristics of included patients with trisomy 8.

Study (Author, year)	Country	Age	Sex	OU	GU	Skin	Joint	GI	Uveitis	Fever	Lung	Vasculitis	MDS	Others	HLA-B51
Cohen Alexa J, 2023 [1]	USA	65	Female	0	0	1	0	0	NA	0	NA	0	1		NA
Sato Yuki, 2023 [2]	Japan	58	Female	1	1	NA	NA	1	1	NA	1*	NA	1		1
Pan, Fei-Yan, 2023 [3]	China	27	Male	NA	NA	NA	NA	NA	NA	1	1**	NA	0		NA
Ding Y, 2023 [4]	China	41	Female	1	1	0	1	0	0	1	NA	NA	1		NA
	China	22	Female	1	1	0	0	0	0	0	NA	0	1		NA
	China	56	Female	1	0	1	1	0	0	0	NA	0	1		NA
	China	38	Female	1	1	0	1	1	0	1	NA	0	1		NA
	China	75	Male	1	1	0	0	1	0	1	NA	0	1		NA
	China	43	Male	1	1	0	0	1	0	0	NA	0	1		NA
	China	48	Female	1	1	0	0	1	0	1	NA	0	1		NA
	China	37	Female	1	1	0	1	1	0	0	NA	0	1		NA
	China	58	Female	1	0	0	0	1	0	1	NA	0	1		NA
	China	64	Male	1	1	0	0	1	0	1	NA	0	1		NA
	China	36	Female	1	0	1	0	1	0	1	NA	0	1		NA
Ishikawa Y, 2023 [5]	Japan	82	Male	NA	NA	1	1	1	NA	1	NA	NA	1		NA
Takahashi N, 2023 [6]	Japan	67	Male	0	0	1	NA	0	0	1	NA	0	1		0
Takahashi R 2023 [7]	Japan	80	Male	1	0	0	NA	1	NA	1	NA	NA	1		0
Onaka T, 2022 [8]	Japan	49	Female	1	1	0	1	1	0	1	NA	NA	1		0
	Japan	52	Female	1	1	1	NA	1	0	1	NA	NA	0		1
Mori Y.2022 [9]	Japan	70	Female	NA	0	NA	NA	1	0	0	NA	NA	0		0
Buchrits Shira, 2022 [10]	Israel	57	Male	NA	NA	1	1	NA	0	1	NA	1	0	AML	NA
	Israel	43	Male	NA	NA	NA	1	NA	0	1	NA	0	0	AML	NA
	Israel	24	Male	NA	NA	1	NA	NA	0	NA	NA	0	0	AML	NA
Fu YK, 2022 [11]	China	69	Male	0	NA	0	1	0	NA	1	NA	NA	0		NA
	China	36	Male	0	NA	0	1	1	0	1	NA	NA	0		NA
	China	67	Male	0	NA	0	1	0	NA	1	NA	NA	0		NA
	China	62	Male	0	NA	1	1	0	NA	1	NA	NA	0		NA
	China	66	Male	1	NA	0	0	1	NA	1	NA	NA	0		NA
	China	40	Female	0	NA	1	0	1	NA	1	NA	NA	0		NA
Xu Guan-Hua, 2022 [12]	China	18	Male	NA	NA	NA	0	NA	0	1	NA	NA	1		NA
Shimizu H, 2021 [13]	Japan	58	Female	1	1	1	NA	1	0	1	1*	NA	1		1
Zhang X, 2021 [14]	China	63	Male	1	NA	1	NA	1	NA	1	NA	NA	1		NA
Wei Q, 2021 [15]	China	52	Female	1	1	NA	1	1	NA	1	NA	NA	1		NA
Lee S, 2021 [16]	Korea	29	Male	NA	NA	NA	NA	1	NA	1	NA	NA	1		NA
Sun Y, 2021 [17]	USA	83	Female	NA	NA	NA	NA	NA	NA	1	NA	NA	1		NA
Ishii A, 2021 [18]	Japan	45	Female	1	1	1	NA	1	NA	1	NA	NA	1		0
Abhyankar Priya, 2020 [19]	USA	58	Male	1	NA	NA	1	1	NA	NA	NA	NA	1		NA
Paul Shilpa, 2020 [20]	France	26	Male	NA	NA	1	NA	NA	NA	NA	NA	NA	0		NA
	France	57	Female	NA	NA	1	NA	NA	NA	NA	NA	NA	1		NA
	France	61	Female	NA	NA	1	NA	NA	NA	1	NA	NA	0		NA
Mori Y, 2020 [21]	Japan	70	Male	1	0	0	NA	1	0	1	NA	NA	1		1
Oka S, 2020 [22]	Japan	73	Female	1	NA	NA	NA	1	NA	1	NA	NA	1		0
Asano T, 2019 [23]	Japan	18	Female	1	1	NA	1	1	NA	1	NA	NA	1		0
Tanaka N, 2019 [24]	Japan	56	Female	1	NA	NA	NA	1	NA	1	NA	NA	1		NA
Wesner N, 2019 [25]	France	75(65-87), median (range)	Female6 Male5	10	4	3	5	6	0	NA	NA	NA	11		NA
Narazaki T, 2019 [26]	Japan	55	Female	1	0	0	NA	1	1	1	NA	NA	0	PMF	1
Kono M, 2019 [27]	Japan	63	Male	1	0	0	NA	1	NA	1	NA	NA	1		1
Wesner N, 2019 [28]	France	67(59-80), Median (range)	Female11 Male10	10	4	7	12	11	0	NA	NA	2	21		NA
Awaji K, 2018 [29]	Japan	67	Male	1	1	1	1	1	NA	1	NA	NA	0		NA
Shen Y, 2018 [30]	China	49.5±12.8, mean±SD	Female8 Male2	10	8	4	3	9	0	6	NA	NA	10		NA
Chen Y, 2017 [31]	China	46	Female	1	1	NA	1	1	NA	NA	NA	NA	1		NA
Nakamura Y, 2017 [32]	Japan	38	Male	1	1	1	NA	1	0	1	NA	0	0	AML	0
Lee WS, 2017 [33]	Korea	52	Female	1	1	0	NA	1	1	1	NA	NA	1		NA
Nakanishi K, 2016 [34]	Japan	73	Male	NA	NA	1	NA	1	NA	1	NA	NA	0	CML	
Kimura, M 2015 [35]	Japan	79	Female	1	1	NA	NA	1	NA	NA	NA	NA	1		1
Kawano S, 2015 [36]	Japan	81	Male	1	0	0	NA	1	0	NA	NA	NA	1		0
	Japan	45	Male	1	1	NA	NA	1	NA	NA	NA	NA	1		1
	Japan	64	Male	NA	NA	NA	NA	1	NA	NA	NA	NA	1		NA
Soysal T, 2014 [37]	Turkey	30	Female	1	1	1	1	1	NA	1	NA	NA	1		NA
Muñoz-Grajales C, 2014 [38]	Venezuela	24	Female	1	1	1	NA	1	0	1	NA	NA	0	CML	0
Koguchi-Yoshioka, H 2014 [39]	Japan	63	Male	1	1	0	NA	1	NA	1	NA	NA	1		0
Handa T, 2014 [40]	Japan	32	Female	1	1	1		1	NA	NA	1*	NA	1		0
	Japan	26	Male	1	0	1		1	NA	NA	1*	NA	1		0
	Japan	38	Female	1	1	1		0	1	NA	1*	NA	1		NA
Kulasekararaj AG, 2013 [41]	England	70	Male	NA	NA	NA	NA	NA	NA	NA	0	NA	1		NA
Tanaka, H, 2013 [42]	Japan	59	Male	1	1	NA	NA	1	NA	1	NA	NA	1		NA

Study (Author, year)	Country	Age	Sex	OU	GU	Skin	Joint	GI	Uveitis	Fever	Lung	Vasculitis	MDS	Others	HLA-B51
Toyonaga, T. 2013 [43]	Japan	36	Male	1	0	0	NA	1	NA	0	NA	NA	1		1
	Japan	29	Female	1	1	1	NA	1	NA	1	NA	NA	1		NA
Chen HC, 2012 [44]	Taiwan	24	Female	1	1	1	NA	1	NA	1	NA	NA	1		NA
Merrill, A 2011 [45]	USA	53	Female	NA	NA	1	1	NA	NA	1	1***	1****	1		NA
Kovacs E, 2009 [46]	Hungary	75	Male	1	1	NA	1	1	NA	NA	NA	NA	1		NA
Mantzourani MG, 2009 [47]	Greece	68	Male	1	1	1	NA	1	NA	1	NA	NA	0	CMML	0
Nakamura F, 2009 [48]	Japan	55	Male	0	0	0	NA	1	0	1	NA	NA	1		NA
Ahn JK, 2008 [49]	Korea	34	Female	1	1	1	1	1	0	NA	NA	NA	1		NA
	Korea	53	Male	1	1	1	0	1	0	NA	NA	NA	1		NA
	Korea	49	Female	1	1	0	0	1	1	NA	NA	NA	1		NA
	Korea	47	Male	1	1	1	0	0	0	NA	NA	NA	1		NA
	Korea	46	Female	1	1	1	0	1	0	NA	NA	NA	1		NA
	Korea	35	Female	1	1	1	0	1	0	NA	NA	NA	1		NA
	Korea	43	Male	1	1	1	0	0	0	NA	NA	NA	1		NA
Hoetzenecker W, 2008 [50]	Germany	74	Male	NA	NA	1	NA	NA	NA	NA	1*	NA	1		0
Nonami A, 2007 [51]	Japan	28	Female	1	0	0	NA	1	0	1	NA	NA	1		
Fine LA, 2007 [52]	USA	28	Female	1	1	NA	1	1	NA	0	NA	NA	1		1
Koca E, 2006 [53]	Turkey	40	Female	NA	NA	1	NA	NA	NA	NA	NA	NA	1		NA
Kawabata H, 2006 [54]	Japan	76	Female	1	NA	NA	NA	1	NA	1	NA	NA	1		1
	Japan	75	Male	1	1	1	NA	1	NA	1	NA	NA	1		NA
	Japan	67	Female	1	NA	NA	NA	1	NA	1	NA	NA	1		NA
Hamada T, 2008 [55]	Japan	33	Male	0	0	1	0	0	NA	1	0	0	0		NA
Oliveira W, 2009 [56]	Brazil	19	Male	0	0	0	0	0	NA	1	0	0	1		NA
Yano K, 1996 [57]	Japan	23	Female	1	1	1	0	1	0	0	0	0	1		0
	Japan	54	Female	1	1	1	0	0	0	1	0	0	1		NA
Billström R, 1997 [58]	Sweden	54	Male	1	0	1	0	0	NA	1	1	1	1		NA
Ohno E, 1997 [59]	Japan	34	Female	1	1	0	1	1	1	1	0	0	1		0
Kuroda J, 2000 [60]	Japan	74	Male	NA	NA	NA	NA	NA	NA	1	1	NA	1		NA
Kimura S, 2001 [61]	Japan	74	Female	1	NA	NA	NA	1	NA	NA	NA	NA	1		NA
	Japan	36	Female	NA	NA	NA	NA	NA	NA	1	NA	NA	1		NA
	Japan	31	Female	NA	NA	NA	NA	1	NA	NA	NA	NA	1		NA
Apostolou T, 2002 [62]	Greece	61	Male	NA	NA	1	NA	NA	NA	NA	NA	NA	1		NA
Adachi Y, 2003 [63]	Japan	28	Female	1	1	1	NA	1	0	1	NA	NA	1		0
	Japan	39	Female	1	1	1	NA	1	0	NA	NA	NA	1		0
Hattori H, 2003 [64]	Japan	69	Male	NA	NA	1	NA	0	NA	1	NA	NA	1		NA
Hasegawa H, 2003 [65]	Japan	56	Female	1	1	1	1	NA	NA	1	0	NA	1		0
Kataoka K, 2009 [66]	Japan	76	Female	NA	NA	NA	NA	NA	NA	1	1	NA	1		NA
Fujimura T, 2010 [67]	Japan	74	Male	NA	NA	1	NA	0	NA	1	NA	NA	1		NA
	Japan	71	Male	1	0	1	0	0	0	1	NA	NA	1		NA
Nishida A, 2011 [68]	Japan	35	Male	0	0	0	0	1	NA	0	1	NA	1		NA
	Japan	42	Female	0	0	0	0	1	NA	0	1	NA	1		NA
	Japan	58	Female	NA	NA	NA	NA	1	NA	1	1	NA	1		NA
Nishida A, 2011 [69]	Japan	47	Male	NA	NA	NA	NA	NA	NA	NA	1	NA	1		NA
Kaffenberger B, 2012 [70]	USA	60	Female	NA	NA	1	NA	NA	NA	1	NA	NA	1		NA
Nishikawa Y, 2013 [71]	Japan	65	Male	NA	NA	1	NA	NA	NA	1	NA	NA	1		NA
Kook MH, 2014 [72]	Korea	38	Female	1	1	NA	1	1	1	1	NA	NA	1		NA
Moriyama M, 2015 [73]	Japan	44	Female	NA	NA	NA	NA	NA	NA	NA	1	NA	1		NA
Endo M, 2015 [74]	Japan	68	Female	1	0	NA	0	1	0	1	NA	NA	1		0
Haga N, 2016 [75]	Japan	87	Male	1	1	1	NA	1	NA	NA	NA	NA	1		0
Ito K, 2016 [76]	Japan	32	Male	1	1	NA	NA	NA	NA	1	NA	NA	1		NA
	Japan	58	Male	NA	NA	NA	NA	1	NA	NA	NA	NA	1		NA
Fukuchi K, 2018 [77]	Japan	85	Male	NA	NA	NA	NA	NA	NA	NA	NA	NA	1		NA
Leonard P, 2022 [78]	Australia	60	Male	1	1	NA	1	1	NA	1	NA	NA	0		NA
Mirouse A, 2014 [79]	France	68	Male	NA	NA	NA	1	NA	NA	1	NA	NA	1		NA
Fujiwara D, 2017 [80]	Japan	47	Female	NA	NA	1	NA	NA	NA	1	NA	0	1		0
Hojo N, 2004 [81]	Japan	73	Male	NA	NA	1	NA	NA	NA	1	NA	NA	1		NA
Yilmaz U, 2020 [82]	Turkey	67	Male	0	0	0	0	1	0	1	NA	NA	1		1
Lin YC, 2008 [83]	China	49	Female	1	1	0	1	1	0	1	NA	1	0	CMML	NA
Kuttikat A, 2005 [84]	UK	54	Male	1	1	1	1	NA	1	NA	NA	NA	1		0
Ogawa H, 2001 [85]	Japan	25	Male	1	1	1	NA	1	0	NA	NA	NA	1		NA
	Japan	41	Female	1	1	1	NA	1	NA	1	NA	NA	1		0
Itamiya T, 2018 [86]	Japan	55	Female	1	0	1	1	1	0	1	NA	NA	1		NA
Liu Z, 2022 [87]	China	40.5 (35.4,51.4), median(IQR)	Female11 Male2	12	5	3	4	13	0	11	NA	NA	13		NA
Sahoo D, 2010 [88]	USA	42	Male	NA	NA	NA	1	NA	NA	1	1	NA	1		NA
Current series, Patient 1	Japan	61	Male	0	0	1	1	0	0	1	1	0	0		NA
Current series, Patient 2	Japan	67	Male	0	0	1	1	1	0	1	0	0	0		NA
Current series, Patient 3	Japan	79	Male	0	0	1	1	0	0	1	1	0	1		NA
Current series, Patient 4	Japan	75	Male	1	1	1	0	1	0	1	0	0	0		NA
Current series, Patient 5	Japan	83	Female	1	0	1	0	1	0	1	0	0	1		0
Current series, Patient 6	Japan	76	Male	1	0	1	1	0	0	1	1	0	1		NA

OU: oral ulcer, GU: genital ulcer, GI: gastrointestinal involvement, MDS: myelodysplastic syndrome, AML: acute myeloid leukemia, CML: Chronic myelogenous leukemia, CMML: chronic myelomonocytic leukemia, PMF: primary myelofibrosis, IQR: Interquartile range, NA: not available.

*: pulmonary alveolar proteinosis, **: pneumonia, ***: acute fibrinous and organizing pneumonia, ****: GCA: giant cell arteritis.

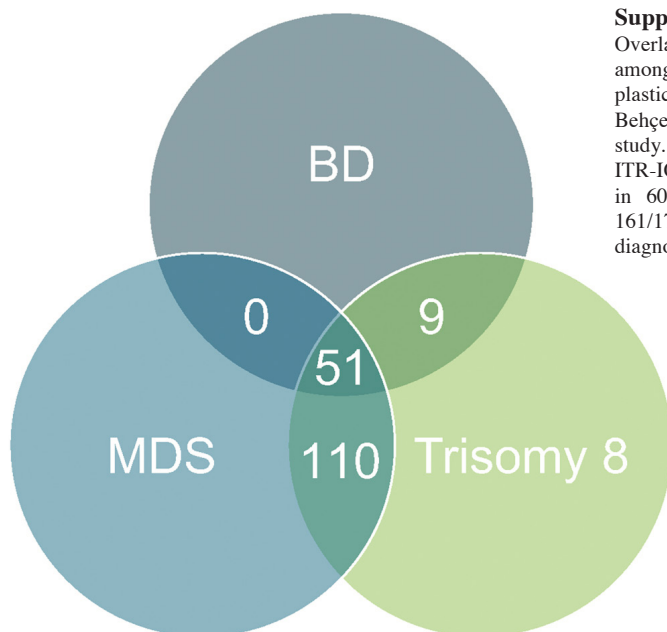
Supplementary Table S2: treatment and outcome of included patients with trisomy 8.

Study (Author, year)	COL	CS	5-ASA	SASP	IS	Bio	JAKi	Chemo	BMT	PBSCT	CBT	Surgery	Others	Outcome	Follow-up time	Death	Cause of death
Cohen Alexa J, 2023 [1]	0	1	0	0	0	0	0	0	0	0	0	0		Improved/Death	1 months	1	Cerebral hemorrhage
Sato Yuki, 2023 [2]	1	1	0	0	1	1	0	0	1	0	0	0		Improved/Stable	> 6 years	0	
Pan, Fei-Yan, 2023 [3]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	22 days	0	
Ding Y, 2023 [4]	0	1	0	0	0	0	0	0	0	0	0	0	Stanozolol, Testosterone undecanoate	Improved/Stable	7 months (n=9)	0	MDS (n=8), severe infection (n=1)
	0	1	0	0	0	0	0	1	0	0	0	0		Improved/Death	NA	1	NA
	0	1	0	0	0	0	0	1	0	0	0	0		Improved/Stable	NA	0	
	0	1	0	0	0	0	0	1	0	0	0	0		Improved/Death	NA	1	NA
	0	1	0	0	1	0	0	1	0	0	0	0		Deteriorated/Death	NA	1	NA
	0	1	0	0	0	0	0	1	0	0	0	0		Deteriorated /Death	NA	1	NA
	0	1	0	1	0	0	0	1	0	0	0	0		Unchanged/Death	NA	1	NA
	0	1	0	0	1	0	0	1	0	0	0	0		Improved/Stable	NA	0	
	0	1	1	0	0	0	0	1	0	0	0	0		Deteriorated/Death	NA	1	NA
	0	1	1	0	0	0	0	0	0	0	0	0		Unchanged/Death	NA	1	NA
	0	1	0	0	1	1	0	1	0	0	0	0	IVIg	Improved/Stable	NA	0	
Ishikawa Y, 2023 [5]	0	0	0	0	0	1	0	0	0	0	0	0		Improved/Death	3.5 years	1	Diverticular hemorrhage
Takahashi N, 2023 [6]	1	1	0	0	0	0	0	0	0	0	0	0		Unchanged	42 months	0	
Takahashi R 2023 [7]	1	1	1	0	0	0	0	1	0	0	0	0	TPN	Improved	1 year	0	
Onaka T, 2022 [8]	0	0	0	0	0	0	0	1	0	1	0	0		Improved	>400 days	0	
	0	0	0	0	0	0	0	1	0	0	1	0		Improved	>500 days	0	
Mori Y, 2022 [9]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	5 months	0	
Buchrits Shira, 2022 [10]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	7 months	0	
	0	1	0	0	0	0	0	0	0	0	0	0		Improved	3 months	0	
	0	1	0	0	0	0	0	1	0	0	0	0		Unchanged	2 weeks	0	
Fu YK, 2022 [11]	1	1	0	0	1	0	1	0	0	0	0	0		Improved	22 months	0	
	0	1	0	0	0	0	1	0	0	0	0	0		Improved	6 months	0	
	0	1	0	0	0	0	1	1	0	0	0	0		Improved	8 months	0	
	0	1	0	0	1	0	0	1	0	0	0	0		lost	lost	0	
	0	1	1	0	0	0	1	0	0	0	0	0		Improved	4 months	0	
	0	1	0	0	1	1	0	0	0	0	0	0		Improved	NA	0	
Xu Guan-Hua, 2022 [12]	0	1	0	1	0	1	0	1	0	0	0	0		Improved/Death	6 months	1	Cerebral hemorrhage
Shimizu H, 2021 [13]	1	1	1	0	0	1	0	0	0	0	0	0		Improved/Unchanged	NA	0	
Zhang X, 2021 [14]	0	1	0	0	0	0	0	1	0	0	0	0		Improved	2 months	0	
Wei Q, 2021 [15]	0	0	0	0	0	0	0	1	0	0	0	0		Improved	1 month	0	
Lee S, 2021 [16]	0	0	0	0	0	1	0	0	0	0	0	0		Stable	NA	0	
Sun Y, 2021 [17]	0	1	0	0	0	0	0	1	0	0	0	0		Deteriorated /Death	<1 month	1	Sepsis
Ishii A, 2021 [18]	1	1	1	0	0	0	0	0	1	0	0	0		Improved	1 year	0	
Abhyankar Priya, 2020 [19]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	10 months	0	
Paul Shilpa, 2020 [20]	0	1	0	0	0	0	0	1	0	0	0	0		Improved	1 week	0	
	0	1	0	0	0	0	0	0	0	0	0	0		Improved	7 months	0	
	0	1	0	0	0	0	0	1	0	0	0	0	IVIg	Improved	1 month	0	
Mori Y, 2020 [21]	0	1	0	0	0	0	0	0	0	0	0	1		Improved	5 years	0	
Oka S, 2020 [22]	0	0	0	0	0	0	0	1	0	0	0	0		Improved/Death	3 months	1	Sepsis
Asano T, 2019 [23]	0	1	0	0	0	0	0	0	0	1	0	0		Improved	7 months	0	
Tanaka N, 2019 [24]	0	1	0	0	0	0	0	1	0	0	0	0		Improved	2 years	0	
Wesner N, 2019 [25]	0	0	0	0	0	0	0	9	0	0	0	0		NA	NA	7	
Narazaki T, 2019 [26]	0	1	0	0	0	1	1	1	0	0	0	0		Improved	3 years 4 months	0	
Kono M, 2019 [27]	0	0	0	0	0	1	0	0	0	0	0	0		Improved	8years	0	
Wesner N, 2019 [28]	1	15	0	0	4	1	0	14	0	0	0	0		NA	21 months	12	NA
Awaji K, 2018 [29]	1	0	1	0	0	0	0	0	0	0	0	0		Improved	NA	0	
Shen Y, 2018 [30]	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA		NA	NA	NA	
Chen Y, 2017 [31]	0	1	0	0	1	1	0	1	0	0	0	0		Improved	4 months	0	
Nakamura Y, 2017 [32]	0	1	1	0	1	0	0	1	1	0	0	0		Improved	45 months	0	
Lee WS, 2017 [33]	0	1	0	1	1	0	0	1	0	1	0	0		Improved/Unchanged	4 years 6 months	0	
Nakanishi K, 2016 [34]	0	1	0	0	0	0	0	0	0	0	0	0	TKI	Improved	4 years	0	
Kimura, M 2015 [35]	0	1	0	0	0	1	0	0	0	0	0	1		Improved	7 months	0	
Kawano S, 2015 [36]	0	1	0	0	1	0	0	0	0	0	0	0		Improved/Death	7 months	1	Urinary infection
	0	0	0	0	0	0	0	0	0	1	0	0		Death	9 months	1	Enteritis, septic shock
	0	0	0	0	0	0	0	0	0	0	1	0		Death	NA	1	Pneumonia
Soysal T, 2014 [37]	0	1	0	0	1	1	0	1	1	0	0	0		Improved	6 years 6 months	0	

Study (Author, year)	COL	CS	5-ASA	SASP	IS	Bio	JAKi	Chemo	BMT	PBSCT	CBT	Surgery	Others	Outcome	Follow-up time	Death	Cause of death
Muñoz-Grajales C, 2014 [38]	1	1	0	0	0	0	0	1	0	0	0	0		Improved/Death	1 month	1	Sepsis
Koguchi-Yoshioka, H 2014 [39]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	NA	0	
Handa T, 2014 [40]	0	1	0	0	1	1	0	0	0	0	0	0		Improved/Death Unchanged/Death	17.5 years 7 years	1 1	Pneumonia PAP deterioration and septic shock
Kulasekararaj AG, 2013 [41]	0	0	0	0	0	0	0	0	0	0	0	0	Whole lung lavage	Improved NA	12 years NA	0 NA	
Tanaka, H, 2013 [42]	0	0	0	0	0	1	0	1	0	0	0	0		Improved	1 year 8 months	0	
Toyonaga, T, 2013 [43]	0	0	0	0	0	0	0	0	0	0	0	1		Deteriorated	>1 month	0	
Chen HC, 2012 [44]	0	1	1	0	1	0	0	0	0	0	0	0		Improved/Death Unchanged	11 years 11 months	0 1	Sepsis
Merrill, A 2011 [45]	0	0	0	0	0	0	0	0	0	0	0	1		Deteriorated	>1 year	1	Worsening of the present illness
Kovacs E, 2009 [46]	0	1	0	0	1	0	0	0	0	0	0	0		Improved/Death	>1 year	1	GI bleeding
Mantzourani MG, 2009 [47]	0	1	0	0	0	0	0	1	0	0	0	0		Unchanged/Death	2 months	1	Acute leukemia
Nakamura F, 2009 [48]	0	0	0	0	1	0	0	0	0	0	0	0		Improved	NA	0	
Ahn JK, 2008 [49]	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA		NA	NA	NA	
	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA		NA	NA	NA	
	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA		NA	NA	NA	
	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA		NA	NA	NA	
	0	0	0	0	0	0	0	0	0	0	0	1		NA	NA	NA	
	0	0	0	0	0	0	0	0	1	0	0	0		NA	NA	NA	
	0	0	0	0	0	0	0	0	1	0	0	0		NA	NA	NA	
Hoetzenecker W, 2008 [50]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	3 months	0	
Nonami A, 2007 [51]	0	0	0	0	0	0	0	0	0	0	1	0	TPN	Improved	480 days	0	
Fine LA, 2007 [52]	1	1	0	0	0	0	0	0	0	0	0	0		Improved	NA	0	
Koca E, 2006 [53]	0	1	0	0	0	0	0	1	0	0	0	0	IFN-alpha2	Improved	14 months	0	
Kawabata H, 2006 [54]	0	0	0	1	0	0	0	0	0	0	0	0	Metenolone	Improved	NA	0	
	0	0	0	1	0	0	0	0	0	0	0	0	TPN	Improved	> 7months	0	
	1	0	0	1	0	0	0	0	0	0	0	0		Unchanged/Death	10 months	1	Bacterial and fungal infection
Hamada T, 2008 [55]	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA		NA	NA	NA	
Oliveira W, 2009 [56]	0	0	0	0	0	0	0	0	0	0	0	0	Danazol	NA	NA	0	
Yano K, 1996 [57]	0	1	0	1	0	0	0	0	0	0	0	0		NA	1 year	0	
	0	1	0	0	0	0	0	0	0	0	0	0		Deteriorated	4 months	1	Pneumonia
Billström R, 1997 [58]	0	1	0	0	1	0	0	0	0	0	0	0		Improved	4 months	0	
Ohno E, 1997 [59]	0	0	0	0	0	0	0	0	0	0	0	0		Improved	7 months	0	
Kuroda J, 2000 [60]	0	1	0	0	0	0	0	0	0	0	0	0		NA	Several months	0	Fungal sepsis
Kimura S, 2001 [61]	0	1	0	0	0	0	0	0	0	0	0	0		NA	6 months	1	
	0	1	0	0	0	0	0	0	0	0	0	0		NA	5 years	0	
	0	1	0	1	0	0	0	0	0	0	0	0		NA	5 years	0	
Apostolou T, 2002 [62]	0	1	0	0	0	0	0	0	0	0	0	0	EPO	NA	1 year	0	
Adachi Y, 2003 [63]	0	1	0	1	1	0	0	0	0	0	0	0		Improved	NA	NA	
	0	1	1	0	0	0	0	0	0	0	0	0		Improved	NA	NA	
Hattori H, 2003 [64]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	5 months	0	
Hasegawa H, 2003 [65]	1	1	0	0	1	0	0	0	0	0	0	0		Unchanged	13 years	0	
Kataoka K, 2009 [66]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	NA	NA	
Fujimura T, 2010 [67]	1	1	0	0	0	0	0	0	0	0	0	0		Improved	1 month	0	
	0	1	0	0	0	0	0	0	0	0	0	0		Improved	3 years	0	
Nishida A, 2011 [68]	0	0	0	0	0	0	0	0	0	1	0	0		Improved	NA	0	
	0	0	0	0	0	0	0	0	0	0	1	0		Deteriorated/Death	NA	1	Pneumonia
	0	0	0	0	0	0	0	0	1	0	0	0		Improved	NA	0	
Nishida A, 2011 [69]	0	0	0	0	0	0	0	0	0	1	0	0		Improved	42 days	0	
Kaffenberger B, 2012 [70]	0	0	0	0	0	0	0	0	0	0	0	0	EPO	Unchanged	1 year	0	
Nishikawa Y, 2013 [71]	0	0	0	0	0	0	0	1	0	0	0	0	Carotene	Improved	2.5 years	0	
Kook MH, 2014 [72]	0	1	0	1	1	0	0	0	0	1	0	0		Improved	229 days	0	
Moriyama M, 2015 [73]	0	0	0	0	0	0	0	0	0	0	1	0		Unchanged/Death	137 days	1	PAP deterioration and respiratory failure
Endo M, 2015 [74]	0	1	0	0	0	0	0	1	0	0	0	0		Improved	10 months	0	

Study (Author, year)	COL	CS	5-ASA	SASP	IS	Bio	JAKi	Chemo	BMT	PBSCT	CBT	Surgery	Others	Outcome	Follow-up time	Death	Cause of death
Haga N, 2016 [75]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	NA	0	
Ito K, 2016 [76]	0	0	0	0	1	0	0	1	0	0	0	0		Unchanged	NA	0	
	0	1	0	0	0	0	0	0	0	0	0	0		Improved	NA	0	
Fukuchi K, 2018 [77]	0	1	0	0	1	0	0	0	0	0	0	0		Improved	NA	0	
Leonard P, 2022 [78]	0	1	0	0	0	1	0	0	0	0	0	0	zinc	Improved	several months	0	
Mirouse A, 2014 [79]	0	1	0	0	0	0	0	1	0	0	0	0		Improved	6 months	0	
Fujiwara D, 2017 [80]	0	1	0	0	0	0	0	0	0	1	0	0	DDS	Improved	3 months	0	
Hojo N, 2004 [81]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	NA	0	
Yilmaz U, 2020 [82]	0	1	1	0	0	1	0	1	0	0	0	0		Improved	10 months	1	Aspiration pneumonia
Lin YC, 2008 [83]	0	1	0	0	0	0	0	0	0	0	0	0	IVIg heparin	Unchanged/Death	3 years	1	Septic shock
Kutitkat A, 2005 [84]	0	1	0	0	1	0	0	1	0	0	0	0		Unchanged	NA	0	
Ogawa H, 2001 [85]	0	1	0	0	0	0	0	0	0	0	0	0		Improved	3 years	0	
	0	1	0	0	0	0	0	0	0	0	0	0		Improved	7 months	0	
Itamiya T, 2018 [86]	0	0	0	0	1	0	0	0	0	0	0	0		Improved	NA	0	
Liu Z, 2022 [87]	0	12	5	0	12	0	0	1	0	0	0	0		NA	NA	5	
Sahoo D, 2010 [88]	0	0	0	0	0	0	0	0	0	0	0	0		Unchanged	NA	1	MDS-driven immunologic reaction, MOF
Current series, Patient 1	0	1	0	0	1	0	0	0	0	0	0	0		NA	3 years	0	
Current series, Patient 2	1	1	1	0	0	0	0	0	0	0	0	0		improved	6 months	0	
Current series, Patient 3	1	1	0	0	0	0	0	0	0	0	0	0		improved	4 years	0	
Current series, Patient 4	0	1	1	0	0	1	0	0	0	0	0	0		Deteriorated /Death	8 months	1	Intestinal bleeding
Current series, Patient 5	0	1	1	0	1	1	0	0	0	0	0	0		Improved	5 years	0	
Current series, Patient 6	0	1	0	0	0	0	0	0	0	0	0	0		Deteriorated /Death	5 years	1	MDS overt

COL: colchicine, CS: corticosteroid, 5-ASA: 5-aminosalicylic acid, SASP: salazosulfapyridine, IS: other immunosuppressants, Bio: biologics, Chemo: chemotherapy other than the above, BMT: bone marrow transplantation, PBSCT: peripheral blood stem cell transplantation, CBT: cord blood transplantation.



Supplementary Fig. S3.

Overlap and distinctions among trisomy 8, myelodysplastic syndromes (MDS), and Behçet's disease (BD) in this study.

ITR-ICBD criteria were met in 60/77 cases (77.9%) and 161/179 cases (89.9%) had a diagnosis of MDS.

Supplementary Table S3: laboratory data of included patients with trisomy 8.

Study (Author, year)	WBC (10 ³ /μL)	HB (g/dL)	MCV (fL)	Plt (10 ⁴ /μL)	CRP (mg/dL)
Cohen Alexa J, 2023 [1]	17.2	7	98	14.2	19.82
Sato Yuki, 2023 [2]	NA	NA	NA	NA	NA
Pan, Fei-Yan, 2023 [3]	14.63	10.7	NA	9.8	4.763
Ding Y, 2023 [4]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Ishikawa Y, 2023 [5]	6.7	9.7	NA	7.2	8.25
Takahashi N, 2023 [6]	7.7	7.7	114.9	13.5	22.34
Takahashi R 2023 [7]	4.47	9.4	NA	9.6	13.31
Onaka T, 2022 [8]	2.6	9.4	97.6	9.5	1
	8.6	8.3	70.3	8.7	1
Mori Y.2022 [9]	13.27	10.5	101.791	83.1	2.68
Buchrits Shira, 2022 [10]	NA	8	NA	1.6	NA
	NA	10	NA	8	NA
	NA	7.7	NA	5.3	NA
Fu YK, 2022 [11]	5.17	9.2	112.8	7.6	13.96
	3.22	6.6	120.1	47.8	10.174
	3.77	11.7	116	13.1	1.834
	6.11	7.9	127	12	16.566
	4.89	6.3	108.2	7.9	15
	3.04	10	111.2	23.2	19.49
Xu Guan-Hua, 2022 [12]	3.8	4	122.7	NA	NA
Shimizu H, 2021 [13]	4.8	9.3	NA	14.3	0.35
Zhang X, 2021 [14]	13	8.2	NA	9.3	24.7
Wei Q, 2021 [15]	3.83	9.1	NA	30.4	1.223
Lee S, 2021 [16]	1.7	8.6	NA	0.33?	NA
Sun Y, 2021 [17]	16	8.9/μL?	NA	2.6	NA
Ishii A, 2021 [18]	7.7	6.3	90.5	4.6	14.1
Abhyankar Priya, 2020 [19]	NA	NA	NA	NA	NA
Paul Shilpa, 2020 [20]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Mori Y, 2020 [21]	5.68	8	106.383	10.9	9.03
Oka S, 2020 [22]	1.8	5.7	NA	5.3	2.49
Asano T, 2019 [23]	6.5	13.7	109.7	23.1	0.59
Tanaka N, 2019 [24]	1.49	9.1	95	26.8	2.163
Wesner N, 2019 [25]	NA	NA	NA	NA	NA
Narazaki T, 2019 [26]	15.43	6.8	NA	10.9	NA
Kono M, 2019 [27]	2.9	8.4	101.476	11.7	8.7
Wesner N, 2019 [28]	NA	10(8.7-10.4)	NA	13.7(6.6-30.0)	3.6(0.8-9.7)
Awaji K, 2018 [29]	15.5	11.7	NA	NA	NA
Shen Y, 2018 [30]	3.7±2.5	8.97±1.77	NA	16.39±11.64	6.33±4.14
Chen Y, 2017 [31]	2.8	10.7	NA	NA	10.45
Nakamura Y, 2017 [32]	10.4	8.6	NA	7.1	NA
Lee WS, 2017 [33]	2.08	7.5	NA	7.2	NA
Nakanishi K, 2016 [34]	32.3	13.8	NA	13.7	30.05
Kimura, M 2015 [35]	1.8	5.9	NA	NA	18.4
Kawano S, 2015 [36]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Soysal T, 2014 [37]	NA	NA	NA	NA	NA
Muñoz-Grajales C, 2014 [38]	NA	NA	NA	NA	NA
Koguchi-Yoshioka, H 2014 [39]	NA	7.7	114.8	NA	17.9
Handa T, 2014 [40]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Kulasekararaj AG, 2013 [41]	4.66	12.4	NA	21.5	NA
Tanaka, H, 2013 [42]	3.8	8.3	NA	11.1	2.34
Toyonaga, T. 2013 [43]	2.5	11.2	92.3	NA	10.3
	NA	NA	NA	NA	NA

Study (Author, year)	WBC (10 ³ /μL)	HB (g/dL)	MCV (fL)	Plt (10 ⁴ /μL)	CRP (mg/dL)
Chen HC, 2012 [44]	12.5	14.1	NA	15	NA
Merrill, A 2011 [45]	NA	NA	108	NA	NA
Kovacs E, 2009 [46]	4.95	8.1	NA	8	NA
Mantzourani MG, 2009 [47]	16.26	12.1	NA	18.4	13.1
Nakamura F, 2009 [48]	2.1	9.5	123.2	9.2	16.9
Ahn JK, 2008 [49]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Hoetzenecker W, 2008 [50]	11.2	8.6	NA	124	NA
Nonami A, 2007 [51]	1.5	11	NA	7	NA
Fine LA, 2007 [52]	3.3	NA	108	14.5	NA
Koca E, 2006 [53]	58	7	NA	NA	NA
Kawabata H, 2006 [54]	NA	5.7	101.1	NA	18
	16.6	12.2	NA	16.6	16.7
	NA	NA	NA	NA	NA
Hamada T, 2008 [55]	7.4	10.8	NA	27.1	3.4
Oliveira W, 2009 [56]	1.5	8	NA	1.5	NA
Yano K, 1996 [57]	NA	9.1	NA	6.1	12.7
	6.5	6.1	NA	NA	NA
Billström R, 1997 [58]	2.7	11.8	123	NA	NA
Ohno E, 1997 [59]	6.22	9.1	NA	4.2	43.5
Kuroda J, 2000 [60]	6.15	12.3	123	NA	NA
Kimura S, 2001 [61]	NA	NA	NA	NA	6.2
	NA	NA	NA	NA	26.1
	NA	NA	NA	NA	NA
Apostolou T, 2002 [62]	5.5	8	NA	17	NA
Adachi Y, 2003 [63]	2.6	5.4	NA	0.9	NA
	2	5.4	NA	0.7	NA
Hattori H, 2003 [64]	9.29	8.9	110.5	8.4	9
Hasegawa H, 2003 [65]	4.9	11.4	99.7	19.2	15.62
Kataoka K, 2009 [66]	5.1	8.7	NA	37.8	7.24
Fujimura T, 2010 [67]	18	7.5	105.8	22.5	14.3
	23.7	95	114.1	3.4	38.1
Nishida A, 2011 [68]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Nishida A, 2011 [69]	NA	NA	NA	NA	NA
Kaffenberger B, 2012 [70]	NA	NA	NA	NA	NA
Nishikawa Y, 2013 [71]	9.9	6.2	NA	0.4	NA
Kook MH, 2014 [72]	1.7	5.1	NA	9	NA
Moriyama M, 2015 [73]	NA	NA	NA	NA	NA
Endo M, 2015 [74]	NA	9.9	89.9	6	31.7
Haga N, 2016 [75]	NA	9.3	118	NA	7.81
Ito K, 2016 [76]	NA	NA	NA	NA	NA
	NA	NA	NA	NA	NA
Fukuchi K, 2018 [77]	29.44	13.4	NA	NA	NA
Leonard P, 2022 [78]	NA	NA	NA	NA	14
Mirouse A, 2014 [79]	NA	7.1	NA	NA	5.5
Fujiwara D, 2017 [80]	18.5	9.1	NA	4.9	19.67
Hojo N, 2004 [81]	8.1	6.2	78.9	13.4	32.8
Yilmaz U, 2020 [82]	5.8	9.1	126	18.5	7.88
Lin YC, 2008 [83]	30.6	14.1	NA	24.3	NA
Kuttikat A, 2005 [84]	NA	12	103	NA	NA
Ogawa H, 2001 [85]	NA	11.6	NA	2.5	1
	NA	11.5	NA	6	17.3
Itamiya T, 2018 [86]	2.2	6.4	NA	5.7	7.04
Liu Z, 2022 [87]	2.4	6.6	NA	6.2	5.79
Sahoo D, 2010 [88]	NA	NA	NA	NA	NA
Current series, Patient 1	NA	NA	NA	NA	NA
Current series, Patient 2	3.1	9.8	90.3	18.5	0.07
Current series, Patient 3	4.46	13.4	105	10.8	0.04
Current series, Patient 4	3.2	7.5	97.4	19.5	3.36
Current series, Patient 5	4.9	9	90.7	7.7	0.21
Current series, Patient 6	11.5	10.9	118.4	19.6	13.9

References

- COHEN AJ, GLEASON LK, BHATTI SA, NIK-BAKHT N: Neutrophilic panniculitis associated with myelodysplastic syndrome/myeloproliferative neoplasm: a case report and literature review. *Int J Dermatol Venereol* 2023; 6(3): 168-71. <https://doi.org/10.1097/jd9.0000000000000286>
- SATO Y, FUKATSU M, SUZUKI T *et al.*: Successful allogeneic hematopoietic stem cell transplantation for myelodysplastic neoplasms complicated with secondary pulmonary alveolar proteinosis and Behçet's disease harboring GATA2 mutation. *Int J Hematol* 2023; 118(5): 642-6. <https://doi.org/10.1007/s12185-023-03603-0>
- PAN FY, FAN HZ, ZHUANG SH, PAN LF, YE XH, TONG HJ: Severe inflammatory disorder in trisomy 8 without myelodysplastic syndrome and response to methylprednisolone: A case report. *World J Clin Cases* 2023; 11(26): 6206-12. <https://doi.org/10.12998/wjcc.v11.i26.6206>
- DING Y, HU W, LI L *et al.*: Clinical features and independent predictors of Behçet's disease associated with myelodysplastic syndrome. *Clin Exp Rheumatol* 2023; 41(9): 1823-30. <https://doi.org/10.55563/clindexrheumatol/04us5e>
- ISHIKAWA Y, SASAKI R, ISHIWATA A, HATAKEYAMA S, MATSUMURA M, SATO T: A case of Behçet's-like disease associated with trisomy 8-positive myelodysplastic syndrome carrying MEFV E148Q variant presented with periodic fever. *Mod Rheumatol Case Rep* 2023; 7(2): 470-4. <https://doi.org/10.1093/mrcr/rxad015>
- TAKAHASHI N, HANAJIRI R, SUZUKI M *et al.*: <Editors' Choice> Myelodysplastic syndrome with trisomy 8 presenting periodic fever and multiple MEFV gene variants outside exon 10: a case report. *Nagoya J Med Sci* 2023; 85(1): 195-203. <https://doi.org/10.18999/nagjms.85.1.195>
- TAKAHASHI R, MATSUBARA Y, TAKAHASHI S *et al.*: Refractory Intestinal Behçet-Like Disease Associated with Trisomy 8 Myelodysplastic Syndrome Resolved by Parenteral Nutrition. *Case Rep Gastroenterol* 2023; 17(1): 87-93. <https://doi.org/10.1159/000533578>
- ONAKA T, NAKANO K, UEMOTO Y *et al.*: Allogeneic stem cell transplantation for trisomy 8-positive myelodysplastic syndrome or myelodysplastic/myeloproliferative disease with refractory Behçet's disease: Case report and the review of literature. *Mod Rheumatol Case Rep* 2022; 6(2): 273-7. <https://doi.org/10.1093/mrcr/rxac032>
- MORI Y, IWAMOTO F, KUNO T *et al.*: Trisomy 8-positive Polycythemia Vera Complicated with Intestinal Behçet's-like Disease: A New Perspective for a Clinical Approach. *Intern Med* 2022; 61(11): 1713-9. <https://doi.org/10.2169/internalmedicine.8395-21>
- BUCHRITS S, KOZLOVOSKI D, GAFTER-GVILI A *et al.*: Autoimmune and inflammatory manifestations associated with acute myeloid leukemia with Trisomy 8-Case series and review of the literature. *Eur J Haematol* 2022; 108(3): 199-203. <https://doi.org/10.1111/ejh.13725>

11. FU Y, WU W, CHEN Z, GU L, WANG X, YE S: Trisomy 8 Associated Clonal Cytopenia Featured with Acquired Auto-Inflammation and Its Response to JAK Inhibitors. *Front Med* 2022; 9. <https://doi.org/10.3389/fmed.2022.895965>
12. XU GH, LIN J, CHEN WQ: Concurrent ankylosing spondylitis and myelodysplastic syndrome: A case report. *World J Clin Cases* 2022; 10(6): 1929-36. <https://doi.org/10.12998/wjcc.v10.i6.1929>
13. SHIMIZU H, SATO S, SUZUKI T *et al.*: Intestinal Behçet's disease complicated by myelodysplastic syndrome and secondary pulmonary alveolar proteinosis: a case report. *BMC Gastroenterol* 2021; 21(1): 488. <https://doi.org/10.1186/s12876-021-02065-0>
14. ZHANG X, JIN M, SHEN Z *et al.*: Intestinal ulcers in a patient with myelodysplastic syndrome: a case report. *BMC Gastroenterol* 2021; 21(1): 372. <https://doi.org/10.1186/s12876-021-01932-0>
15. WEI Q, ZHANG X, PENG Y *et al.*: Successful treatment by thalidomide therapy of intestinal Behçet's disease associated with trisomy 8 myelodysplastic syndrome. *Rheumatology* 2021; 60(6): e200-e2. <https://doi.org/10.1093/rheumatology/keaa848>
16. LEE S, CHOI WH, KIM JS, LEE KA, JEON SR, KIM HS: Trisomy 8 Associated Behçet's Like Disease. *J Rheum Dis* 2021; 28(2): 107-9. <https://doi.org/10.4078/jrd.2021.28.2.107>
17. SUN Y, BLIEDEN C, MERRITT BY, SOSA R, RIVERO G: Hemophagocytic lymphohistiocytosis and myelodysplastic syndrome: a case report and review of the literature. *J Med Case Rep* 2021; 15(1): 98. <https://doi.org/10.1186/s13256-020-02623-2>
18. ISHII A, TSUKAMOTO S, MISHINA T *et al.*: Successful allogeneic bone marrow transplantation after massive gastrointestinal bleeding in a patient with myelodysplastic syndrome associated with intestinal Behçet-like disease. *Leuk Res Rep* 2021; 16: 100278. <https://doi.org/10.1016/j.lrr.2021.100278>
19. ABHYANKAR P, AHMED W, FISCHER M, ROGERS N: A case of myelodysplastic syndrome-associated intestinal Behçet's successfully treated with infliximab. *Am J Gastroenterol* 2020; 115(0): S1476. <https://doi.org/10.14309/01.ajg.0000713344.25196.f2>
20. PAUL S, JAMMAL N, AKHANE N *et al.*: Atypical cases of necrotizing sweet syndrome in patients with myelodysplastic syndrome and acute myeloid leukaemia. *Br J Haematol* 2020; 191(1). <https://doi.org/10.1111/bjh.16937>
21. MORI Y, IWAMOTO F, ISHIDA Y *et al.*: Long-term outcome after surgery in a patient with intestinal Behçet's disease complicated by myelodysplastic syndrome and trisomy 8. *Intest Res* 2020; 18(4): 469-75. <https://doi.org/10.5217/ir.2019.09141>
22. OKA S, ONO K, NOHAWA M: The acquisition of trisomy 8 associated with Behçet's-like disease in myelodysplastic syndrome. *Leuk Res Rep* 2020; 13: 100196. <https://doi.org/10.1016/j.lrr.2020.100196>
23. ASANO T, SATO S, FURUYA MY *et al.*: Intestinal Behçet disease associated with myelodysplastic syndrome accompanying trisomy 8 successfully treated with abdominal surgery followed by hematopoietic stem cell transplantation: A case report. *Medicine* (Baltimore) 2019; 98(46): e17979. <https://doi.org/10.1097/md.00000000000017979>
24. TANAKA N, SAKURABA H, HIRAGA H *et al.*: Long-term maintenance of the mucosal healing induced by azacitidine therapy in a patient with intestinal Behçet's-like disease accompanied with myelodysplastic syndrome involving trisomy 8. *Immunol Med* 2019; 42(3): 135-41. <https://doi.org/10.1080/25785826.2019.1687251>
25. WESNER N, DREVON L, GUEDON A *et al.*: Gastrointestinal Behçet's-like disease with myelodysplastic neoplasms with trisomy 8: a French case series and literature review. *Leuk Lymphoma* 2019; 60(7): 1782-8. <https://doi.org/10.1080/10428194.2018.1542152>
26. NARAZAKI T, SHIRATSUCHI M, TSUDA M *et al.*: Intestinal Behçet's Disease with Primary Myelofibrosis Involving Trisomy 8. *Acta Haematol* 2019; 142(4): 253-6. <https://doi.org/10.1159/000501019>
27. KONO M, SAKURAI T, OKAMOTO K *et al.*: Usefulness of Ustekinumab for Treating a Case of Myelodysplastic Syndrome-associated Inflammatory Bowel Disease. *Intern Med* 2019; 58(14): 2029-33. <https://doi.org/10.2169/internalmedicine.2495-18>
28. WESNER N, DREVON L, GUEDON A *et al.*: Inflammatory disorders associated with trisomy 8-myelodysplastic syndromes: French retrospective case-control study. *Eur J Haematol* 2019; 102(1): 63-9. <https://doi.org/10.1111/ejh.13174>
29. AWAJI K, INOKUCHI R, MAEHARA H *et al.*: Gastrointestinal Behçet's disease: periodic fever, multiple ulcers and trisomy 8 in elderly man. *QJM* 2018; 111(8): 579-80. <https://doi.org/10.1093/qjmed/hcy041>
30. SHEN Y, MA HF, LUO D, CAI JF, ZOU J, GUAN JL: High Incidence of Gastrointestinal Ulceration and Cytogenetic Aberration of Trisomy 8 as Typical Features of Behçet's Disease Associated with Myelodysplastic Syndrome: A Series of 16 Consecutive Chinese Patients from the Shanghai Behçet's Disease Database and Comparison with the Literature. *Biomed Res Int* 2018; 2018: 8535091. <https://doi.org/10.1155/2018/8535091>
31. CHEN Y, SHEN Y, MA HF *et al.*: Infliximab associated with life-threatening lung infection in a patient with Behçet disease with intestinal and hematopoietic system involvement: A case report. *Medicine* (Baltimore) 2017; 96(50): e9202. <https://doi.org/10.1097/md.00000000000009202>
32. NAKAMURA Y, MATSUGUMA M, TOKUNAGA Y *et al.*: Successful Treatment of Behçet's Disease Associated with Acute Myeloid Leukemia with Myelodysplasia-related Changes Using Azacitidine and Tacrolimus before Allogeneic Hematopoietic Stem Cell Transplantation. *InternMed* 2017; 56(10): 1199-202. <https://doi.org/10.2169/internalmedicine.56.7808>
33. LEE WS, KANG MH, HWANG JH, OH YJ, YOO WH: Allogeneic hematopoietic stem cell transplantation for refractory Behçet's disease with myelodysplastic syndrome: a case report. *Int J Rheum Dis* 2017; 20(11): 1860-1. <https://doi.org/10.1111/1756-185x.12669>
34. NAKANISHI K, KINJO M: Mimicker of necrotizing fasciitis with systemic inflammatory response syndrome: recurrent necrotizing Sweet's syndrome associated with chronic myelogenous leukaemia. *BMJ Case Rep* 2016; 2016: bcr2016214461. <https://doi.org/10.1136/bcr-2016-214461>
35. KIMURA M, TSUJI Y, IWAI M *et al.*: Usefulness of Adalimumab for Treating a Case of Intestinal Behçet's Disease with Trisomy 8 Myelodysplastic Syndrome. *Intest Res* 2015; 13(2): 166-9. <https://doi.org/10.5217/ir.2015.13.2.166>
36. KAWANO S, HIRAOKA S, OKADA H, AKITA M, IWAMURO M, YAMAMOTO K: Clinical Features of Intestinal Behçet's Disease Associated with Myelodysplastic Syndrome and Trisomy 8. *Acta Med Okayama* 2015; 69(6): 365-9. <https://doi.org/10.18926/amo/53912>
37. SOYSAL T, SALIHOĞLU A, ESATOĞLU SN *et al.*: Bone marrow transplantation for Behçet's disease: a case report and systematic review of the literature. *Rheumatology* 2014; 53(6): 1136-41. <https://doi.org/10.1093/rheumatology/ket479>
38. MUNOZ-GRAJALES C, GONZÁLEZ FZ, ARENAS RB *et al.*: Behçet's disease, chronic myelomonocytic leukemia and trisomy 8. *Int J Clin Rheumatol* 2014; 9(2): 115-20.
39. KOGUCHI-YOSHIOKA H, INOKUMA D, KANDA M, KONDO M, KIKUCHI K, SHIMIZU S: Behçet's Disease-like Symptoms Associated with Myelodysplastic Syndrome with Trisomy 8: A Case Report and Review of the Literature. *Acta Derm Venereol* 2014; 94(3): 355-6. <https://doi.org/10.2340/00015555-1706>
40. HANDA T, NAKATSUE T, BABAM, TAKADA T, NAKATA K, ISHII H: Clinical features of three cases with pulmonary alveolar proteinosis secondary to myelodysplastic syndrome developed during the course of Behçet's disease. *Respir Investig* 2014; 52(1): 75-9. <https://doi.org/10.1016/j.resinv.2013.05.005>
41. KULASEKARARAJ AG, KORDASTI S, BASU T, SALISBURY JR, MUFTI GJ, DU VIVIER AWP: Chronic relapsing remitting Sweet syndrome - a harbinger of myelodysplastic syndrome. *Br J Haematol* 2015; 170(5): 649-56. <https://doi.org/10.1111/bjh.13485>
42. TANAKA H, SHIMIZU N, TOUGASAKI E *et al.*: Successful treatment by azacitidine therapy of intestinal Behçet's disease associated with myelodysplastic syndrome. *Int J Hematol* 2013; 97(4): 520-4. <https://doi.org/10.1007/s12185-013-1316-x>
43. TOYONAGA T, NAKASE H, MATSUURA M *et al.*: Refractoriness of Intestinal Behçet's Disease with Myelodysplastic Syndrome Involving Trisomy 8 to Medical Therapies - Our Case Experience and Review of the Literature. *Digestion* 2013; 88(4): 217-21. <https://doi.org/10.1159/000355341>
44. CHEN H, CHIU Y: Large-vessel thrombosis in intestinal Behçet's disease complicated with myelodysplastic syndrome and trisomy 8. *World J Gastroenterol* 2012; 18(10): 1137-40. <https://doi.org/10.3748/wjg.v18.i10.1137>
45. MERRILL AL, SMITH H: Myelodysplastic Syndrome and Autoimmunity: A Case Report of an Unusual Presentation of Myelodysplastic Syndrome. *Case Rep Hematol* 2011; 2011:

- 1-4. <https://doi.org/10.1155/2011/560106>
46. KOVACS E, NEMETH H, TELEK B *et al.*: Behçet's disease in a patient with myelodysplastic syndrome. *Clin Lymphoma Myeloma* 2009; 9(6): 459-61. <https://doi.org/10.3816/clm.2009.n.090>
47. MANTZOURANI M, CHANTZIARA K, THANOPOULOU I, VARIAMI H, VAIPOULOS G, PANGALIS G: Coexistence of Behçet's disease and chronic myelomonocyte leukemia with trisomy 8: a case report and review of the literature. *Clin Exp Rheumatol* 2009; 27 (Suppl. 53): S85-7.
48. NAKAMURA F, WATANABE T, HORI K *et al.*: Simultaneous Occurrence of Inflammatory Bowel Disease and Myelodysplastic Syndrome due to Chromosomal Abnormalities in Bone Marrow Cells. *Digestion* 2009; 79(4): 215-9. <https://doi.org/10.1159/000213486>
49. AHN JK, CHA HS, KOH EM *et al.*: Behçet's disease associated with bone marrow failure in Korean patients: clinical characteristics and the association of intestinal ulceration and trisomy 8. *Rheumatology* 2008; 47(8): 1228-30. <https://doi.org/10.1093/rheumatology/ken162>
50. HOETZENECKER W, ZIPFEL M, GUENOVA E, HOETZENECKER K, RÖCKEN M: Vasculitic leg ulcers in a patient with mixed myelodysplastic and myeloproliferative syndrome. *J Eur Acad Dermatol Venereol* 2008; 22(1): 110-2. <https://doi.org/10.1111/j.1468-3083.2007.02271.x>
51. NONAMI A, TAKENAKA K, SUMIDA C *et al.*: Successful Treatment of Myelodysplastic Syndrome (MDS)-related intestinal Behçet's Disease by Up-front Cord Blood Transplantation. *Intern Med* 2007; 46(20): 1753-6. <https://doi.org/10.2169/internalmedicine.46.0291>
52. FINE LA, HOFFMAN LD, HOFFMAN MD: Aphthous ulcerations associated with trisomy 8-positive myelodysplastic syndrome. *J Am Acad Dermatol* 2007; 57(2 Suppl): S38-41. <https://doi.org/10.1016/j.jaad.2006.10.019>
53. KOCA E, DUMAN A, CETINER D *et al.*: Successful treatment of myelodysplastic syndrome-induced pyoderma gangrenosum. *Neth J Med* 2006; 64(11): 422-4.
54. KAWABATA H, SAWAKI T, KAWANAMI T *et al.*: Myelodysplastic Syndrome Complicated with Inflammatory Intestinal Ulcers: Significance of Trisomy 8. *Intern Med* 2006; 45(22): 1309-14. <https://doi.org/10.2169/internalmedicine.45.1718>
55. HAMADA T, MATSUURA H, OONO T *et al.*: Karyotypic Analysis of Bone Marrow Cells in Pyodermic Lesions Associated with Myelodysplastic Syndrome. *Arch Dermatol* 2008; 144(5): 643-8. <https://doi.org/10.1001/archderm.144.5.643>
56. OLIVEIRA WR, FERREIRA GN, RADY PL, FESTA C, TYRING SK: Epidermodyplasia verruciformis associated with myelodysplastic syndrome: an intriguing association. *J Cutan Med Surg* 2009; 13(6): 317-20.
57. YANO K, EGUCHI K, MIGITAK *et al.*: Behçet's disease complicated with myelodysplastic syndrome a report of two cases and review of the literature. *Clin Rheumatol* 1996; 15(1): 91-3. <https://doi.org/10.1007/bf02231696>
58. BILLSTRÖM R, JOHANSSON B, STRÖMBECK B *et al.*: Clonal CD5-positive B lymphocytes in myelodysplastic syndrome with systemic vasculitis and trisomy 8. *Ann Hematol* 1997; 74(1): 37-40. <https://doi.org/10.1007/s002770050253>
59. OHNO E, OHTSUKA E, WATANABE K *et al.*: Behçet's disease associated with myelodysplastic syndromes. *Cancer* 1997; 79(2): 262-8. [https://doi.org/10.1002/\(sici\)1097-0142\(19970115\)79:2%3C262::aid-cncr9%3E3.0.co;2-m](https://doi.org/10.1002/(sici)1097-0142(19970115)79:2%3C262::aid-cncr9%3E3.0.co;2-m)
60. KURODA J, KIMURA S, AKAOGI T *et al.*: Myelodysplastic Syndrome with Clonal Eosinophilia Accompanied by Eosinophilic Pulmonary Interstitial Infiltration. *Acta Haematol* 2000; 104(2-3): 119-23. <https://doi.org/10.1159/000039744>
61. KIMURA S, KURODA J, AKAOGI T, HAYASHI H, KOBAYASHI Y, KONDO M: Trisomy 8 Involved in Myelodysplastic Syndromes as a Risk Factor for Intestinal Ulcers and Thrombosis - Behçet's Syndrome. *Leuk Lymphoma* 2001; 42(1-2): 115-21. <https://doi.org/10.3109/10428190109097683>
62. APOSTOLOU T, SOTSIOU F, PAPPAS C, RONTIANNI D, APOSTOLIDIS J, NIKOLOPOULOU N: Atheroembolic renal disease and membranous nephropathy, in a patient with myelodysplastic syndrome, eosinophilia, and trisomy 8. *Nephrol Dial Transplant* 2002; 17(7): 1336-8. <https://doi.org/10.1093/ndt/17.7.1336>
63. ADACHI Y, TSUTSUMI A, MURATA H *et al.*: Behçet's disease accompanied by myelodysplastic syndrome with trisomy 8: two case reports and a review of 15 Japanese cases. *Mod Rheumatol* 2003; 13(1): 90-4. <https://doi.org/10.3109/s101650300015>
64. HATTORI H, HOSHIDA S, YONEDA S: Sweet's Syndrome Associated with Recurrent Fever in a Patient with Trisomy 8 Myelodysplastic Syndrome. *Int J Hematol* 2003; 77(4): 383-6. <https://doi.org/10.1007/sf02982648>
65. HASEGAWA H, IWAMASA K, HATTA N, FUJITA S: Behçet's disease associated with myelodysplastic syndrome with elevated levels of inflammatory cytokines. *Mod Rheumatol* 2003; 13(4): 350-5. <https://doi.org/10.3109/s10165-003-0245-6>
66. KATAOKA K, ICHIKAWA M, HANGAISHI A, TAKAHASHI T, IMAI Y, KUROKAWA M: Interstitial pneumonia associated with progression of myelodysplastic syndrome. *Int J Hematol* 2009; 89(5): 718-9. <https://doi.org/10.1007/s12185-009-0328-z>
67. FUJIMURA T, YUKAWA N, NAKASHIMA R *et al.*: Periodic fever and erythema nodosum associated with MDS with trisomy 8: report of two cases and review of the literature. *Mod Rheumatol* 2010; 20(4): 413-9. <https://doi.org/10.1007/s10165-010-0291-9>
68. NISHIDA A, MIYAMOTO A, YAMAMOTO H *et al.*: Possible Association of Trisomy 8 with Secondary Pulmonary Alveolar Proteinosis in Myelodysplastic Syndrome. *Am J Respir Crit Care Med* 2011; 184(2): 279-80. <https://doi.org/10.1164/ajrcm.184.2.279a>
69. NISHIDA A, OTA H, IKEBE T *et al.*: Allogeneic Hematopoietic Stem Cell Transplantation (HSCT) for Secondary Pulmonary Alveolar Proteinosis with Trisomy-8 Myelodysplastic Syndrome. *Blood* 2011; 118(21): 4705. <https://doi.org/10.1182/blood.V118.21.4705.4705>
70. KAFFENBERGER B, DARABI K, PETERS S, KYNKY J, BECHTEL M: Generalized eruptive histiocytomas and Rosai-Dorfman disease presenting concurrently in a patient with myelodysplastic syndrome. *J Clin Aesthet Dermatol* 2012; 5(8): 42-6.
71. NISHIKAWA Y, OKUDA S, TAKEBAYASHI C, TANIMOTO T, KAMI M, KOBAYASHI K: A case of late onset erythropoietic protoporphyria associated with myelodysplastic syndrome treated by the combination of beta carotene and azacitidine. *Ann Hematol* 2013; 92(10): 1415-6. <https://doi.org/10.1007/s00277-013-1720-6>
72. KOOK M-H, YHIM H-Y, LEE N-R *et al.*: Successful treatment of myelodysplastic syndrome and Behçet colitis after allogeneic hematopoietic stem cell transplantation. *Korean J Intern Med* 2014; 29(1): 123. <https://doi.org/10.3904/kjim.2014.29.1.123>
73. MORIYAMA M, YANO T, FURUKAWA T *et al.*: Possible involvement of lung cells harboring an abnormal karyotype in the pathogenesis of pulmonary alveolar proteinosis associated with myelodysplastic syndrome. *Ann Am Thorac Soc* 2015; 12(8): 1251-53. <https://doi.org/10.1513/annalsats.201503-1751e>
74. ENDO M, SEKIKAWA A, TSUMURA T, MARUO T, OSAKI Y: A Case of Myelodysplastic Syndrome with Intestinal Behçet's Disease-Like Symptoms Treated by Prednisolone and Azacitidine. *Am J Case Rep* 2015; 16: 827-31. <https://doi.org/10.12659/ajcr.895431>
75. HAGAN I, IWATA H, YAMAGUCHI Y *et al.*: Mucocutaneous pyoderma gangrenosum due to trisomy 8 neutrophilic infiltrates in a patient with myelodysplastic syndrome. *Br J Dermatol* 2016; 174(1): 239-41. <https://doi.org/10.1111/bjd.14102>
76. ITO K, KUBOTA K: 18F-FDG PET/CT Findings in Two Cases with Myelodysplastic Syndrome Accompanied by Behçet's disease. *Clin Nucl Med* 2016; 41(8): e392-3. <https://doi.org/10.1097/rlu.0000000000001198>
77. FUKUCHI K, TOKURA Y: Papuloerythroderma occurring in a patient with myelodysplastic syndrome. *J Cutaneous Immunol Allergy* 2018; 1(1): 33-4. <https://doi.org/10.1002/cia2.12007>
78. LEONARD P, LOUW A, PRENTICE D, CIRILLO M: Clonal cytopenia of undetermined significance and atypical Behçet's: the importance of zinc. *BMJ Case Rep* 2022; 15(3): e247154. <https://doi.org/10.1136/bcr-2021-247154>
79. MIROUSE A, VIRONE A, GOBERT D *et al.*: Aseptic muscular abscesses associated with myelodysplastic syndrome. *Eur J Dermatol* 2014; 24(6): 696-7. <https://doi.org/10.1684/ejd.2014.2435>
80. FUJIWARA D, HAMADA T, YAMASAKI O *et al.*: Successful use of etoposide for pyoderma gangrenosum associated with myelodysplastic syndrome and trisomy 8: cytokine profiles during treatment. *Eur J Dermatol* 2017; 27(5): 525-27. <https://doi.org/10.1684/ejd.2017.3067>
81. HOJO N, HASEGAWA H, IWAMASA K, HOJO S,

- FUJITA S: A case of Weber-Christian disease associated with myelodysplastic syndrome. *Mod Rheumatol* 2004; 14(1): 73-6. <https://doi.org/10.1007/s10165-003-0270-5>
82. YILMAZ U, AR MC, ESATOGLU SN *et al.*: How to treat myelodysplastic syndrome with clinical features resembling Behçet syndrome: a case-based systematic review. *Ann Hematol* 2020; 99(6): 1193-203. <https://doi.org/10.1007/s00277-020-03951-5>
83. LIN YC, LIANG TH, CHANG HN, LIN JS, LIN HY: Behcet disease associated with myelodysplastic syndrome. *J Clin Rheumatol* 2008; 14(3): 169-74. <https://doi.org/10.1097/rhu.0b013e3181776bde>
84. KUTTIKAT A, HASKARD D, CHAKRAVARTY K: Behçet's disease associated with trisomy 8 in a male Caucasian patient from Great Britain—a case report. *Clin Exp Rheumatol* 2005; 23 (Suppl. 38): S108-9.
85. OGAWA H, KURODA T, INADA M *et al.*: Intestinal Behçet's disease associated with myelodysplastic syndrome with chromosomal trisomy 8 - A report of two cases and a review of the literature. *Hepatogastroenterology* 2001; 48(38): 416-20.
86. ITAMIYA T, ASAKO K, KIKUCHI H, KONO H: Intestinal Behcet's disease associated with myelodysplastic syndrome with 8 trisomy: a case report and review of Japanese literature. *Clin Exp Rheumatol* 2018; 36 (Suppl. 115): S171.
87. LIU Z, YANG C, BAI X *et al.*: Clinical features and prognosis of patients with gastrointestinal Behcet's disease-like syndrome and myelodysplastic syndrome with and without trisomy 8. *Semin Arthritis Rheum* 2022; 55: 152039. <https://doi.org/10.1016/j.semarthrit.2022.152039>
88. SAHOO D, ASHTON RW, BONFIELD TL, FARVER CF, CULVER DA: Pulmonary alveolar proteinosis and organizing pneumonia in a patient with myelodysplastic syndrome. *Am J Respir Crit Care Med* 2010; 181(1): A4509. https://doi.org/10.1164/ajrccm-conference.2010.181.1_MeetingAbstracts.A4509