Leucemia plasmacellulare

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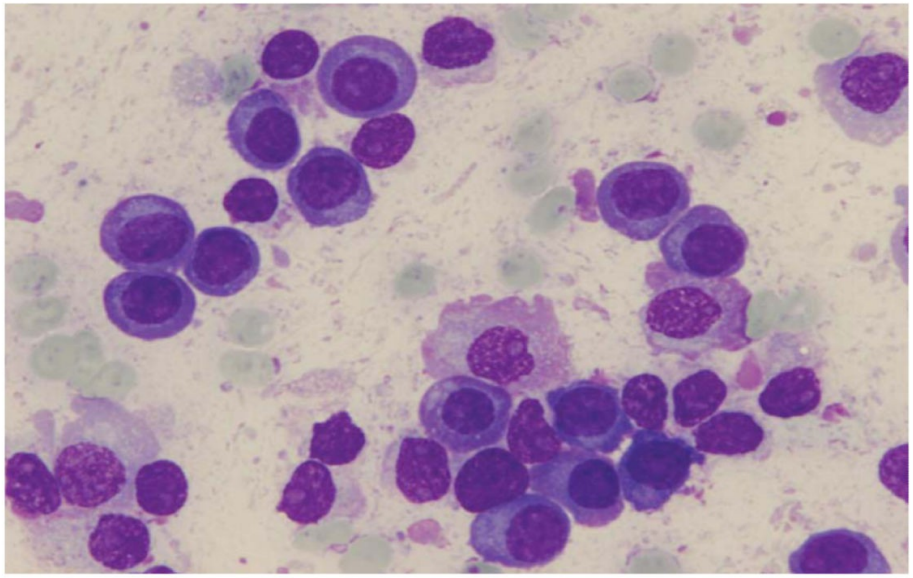
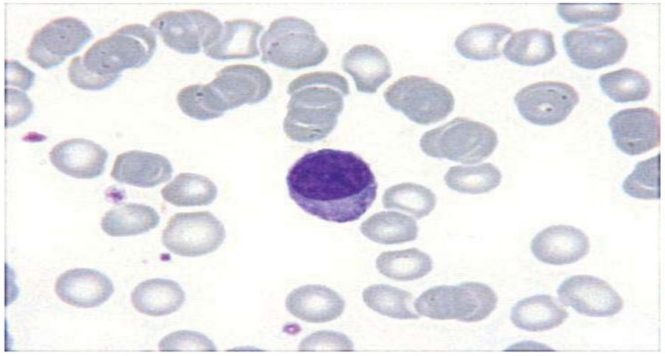
Plasma cell Leukemia (PCL)

- Presence of > 20% plasma cells in peripheral blood
- Absolute plasma cell count > $2 * 10^9/l$
- Primary PCL (pPCL) where PCL is an aspect of primary presentation with no previous myeloma
- Secondary PCL (sPCL): leukemic transformation of relapsed and refractory myeloma

Fernandez de Larrear et al, Leukemia (2013) 27, 780 - 791

A





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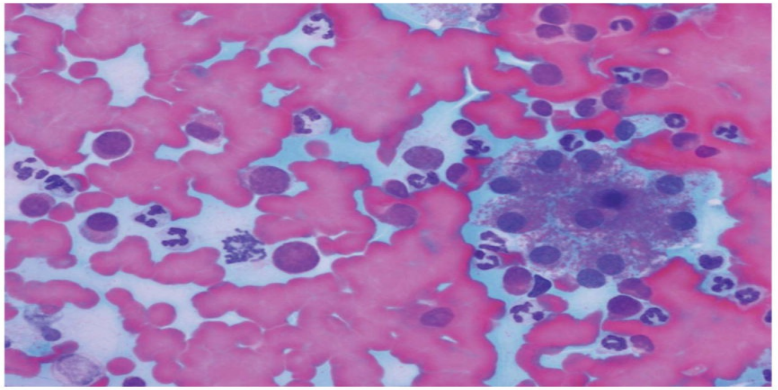

Main clinical and laboratory features of seven retrospective series of patients with primary plasma cell leukemia.

	Noel and Kyle (1987)	Dimopoulos *et al* (1994)	Garcia-Sanz *et al* (1999)	Tiedemann *et al* (2008)	Colovic *et al* (2008)	Peijing *et al* (2009)	Pagano *et al* (2011)
Number of patients	25	27	26	41	30	22	73
Median age (years)	53	57	65	54.5	60	49.5	NA
Sex, M/F	15/10	NA	12/14	24/17	22/8	14/8	43/30
Lytic bone lesions (%)	44	NA	48	35	60	44.4	64
Extramedullary involvement (%)							
Liver	52	32	0	32	56	44.4	23
Spleen	44	18	0	18	53	33.3	18
Lymph nodes	12	6	11	6	3	NA	4
Other	NA	NA	NA	NA	NA	NA	14
M-protein type (%)							
IgG	12.5	52	54	28	53	54.5	30
IgA	25	15	4	13	23	9.1	8
IgD	6	0	8	2	3	0	3
Light chain	44	28	31	41	20	27.3	30
Nonsecretory	12.5	7	4	8	0	9.1	18
Hemoglobin <10 g/dL (%)	>50	82	54	>50	100	>50	48
Platetelet count < 100 × 109/L (%)	>50	67	48	>50	100	>50	NA
High β2-microglobulin (%)	NA	91	65	50	64	50	100
High LDH (%)	NA	63	48	50	37	NA	52
Response to treatment (%)	47	37	38	NA	NA	45.5	55

Cytogenetics of pPCL

Cytogenetics abnormalities (%)	*García-Sanz et al.*[5]	*Dimopoulos et al.*[4]	*Tiedemann et al.*[7]	*Pagano et al.*[13]	*Avet-Loiseau et al.*[51]	*Chiecchio et al.*[52]
Hypodiploidy		41	60	12.2	47	41.6
Hyperdiplody			0	4.9	8.8	33.3
Complex karyotype		92	54.5	34.2	58.8	66.7
del(13q14) or monosomy	84	50	85	19	68	58
del(17p13)			50	7.3	11.8	25
t(11;14)			71	19.5	33	42
t(4;14)			0	0	12	8.3
t(14;16)			0	0	16	25

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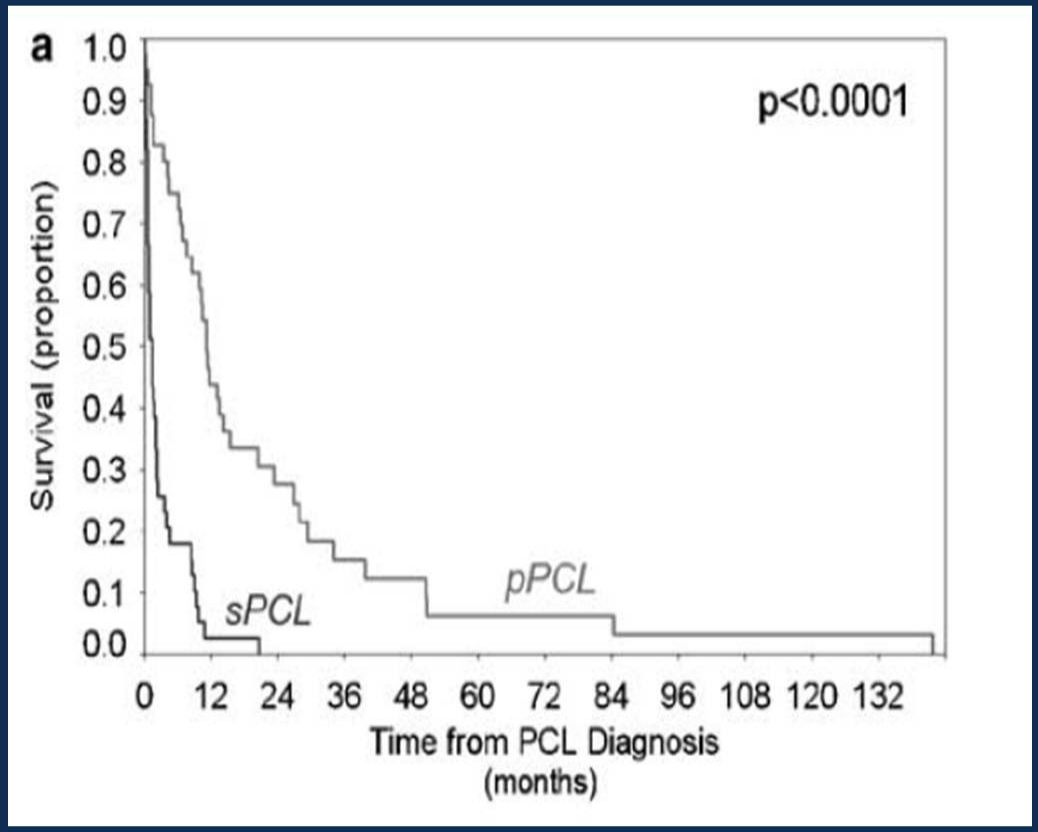
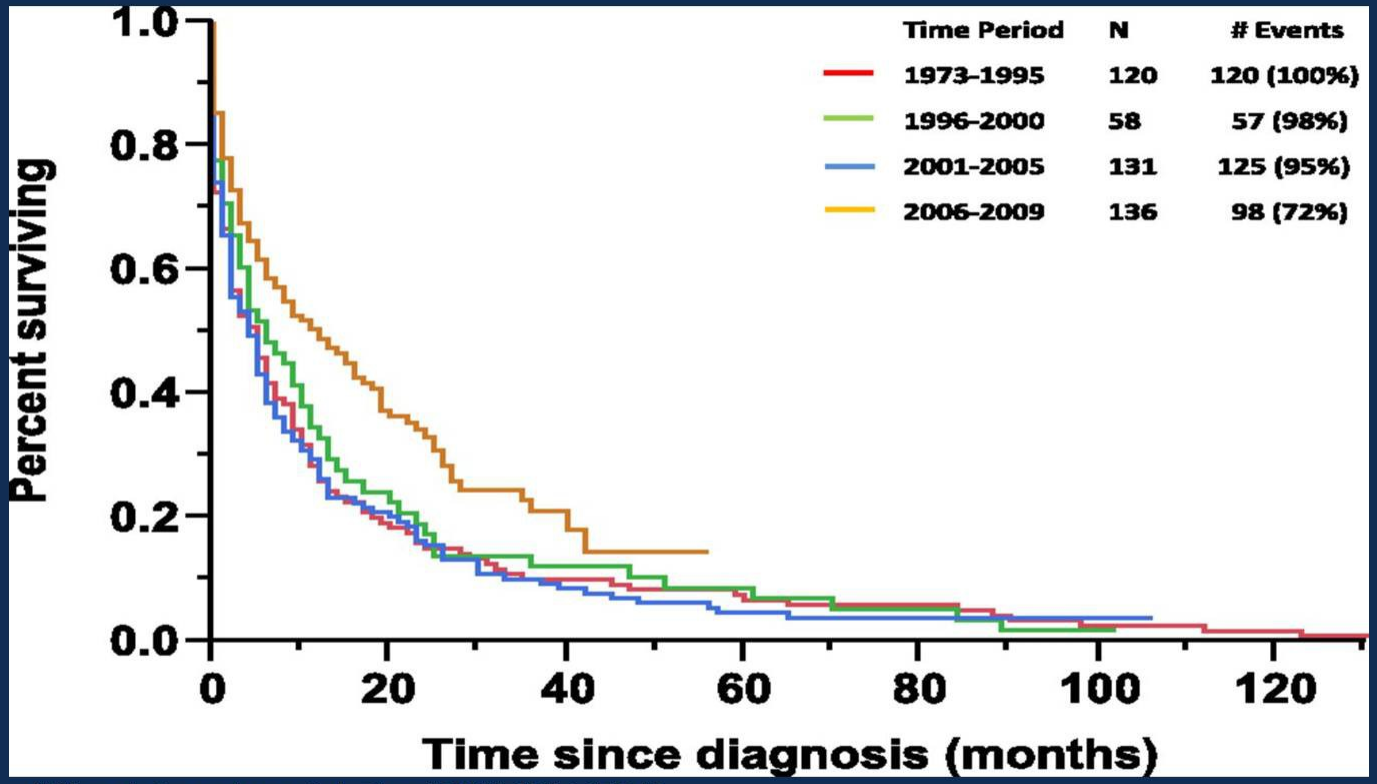
Survival: pPCL vs sPCL



Tiedemann, Leukemia (208) 22, 1044 - 1052

Kaplan-Meier Curve for OS in pPCL patients based on period of diagnosis.



Wilson I. Gonsalves et al. Blood 2014;124:907-912

Treatment options

TARGET THERAPY

Table 3. Results from selected studies on NA in primary PCL patients.

Authors	N of pts	Induction	≥PR after induction	Pts receiving SCT	PFS	OS
RETROSPECTIVE STUDIES						
D'Arena et al. [54]	29	Bortezomib-based	79%	41%	40% @ 2 yr	55% @ 2 yr
Katroditou et al. [55]	25	Bortezomib-based (69%)	80%	24% (overall)	—	50% @ 18 mo
Reece et al. [56]	10	Bortezomib, Cyclophosphamide and dexamethasone	100%	90%	50% @ 18 mo	
PROSPECTIVE STUDIES						
Musto et al. [49]	23	Lenalidomide, Dexamethasone	74%	39%	50% @ 15 mo	50% @ 28 mo (median NR post-SCT)
Royer et al. [57]	40	Bortezomib, doxorubicin and dexamethasone alternating to Cyclophosphamide, bortezomib and dexamethasone	69%	65%	50% @ 16 mo	50% @ 3 yr post-SCT

PFS: progression-free survival; OS: overall survival; SCT: stem cell transplant; pts: patients; mo: months; yr: year; PR: partial response; NA: not available; NR: not reached.

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Autologous and allogenic transplatation

Table 4. Results from selected, retrospective studies with autologous and allogeneic transplantation in PCL patients.

Authors	N of Pts	Preparative regimen	NRM	PFS	OS
AUTOLOGOUS STEM CELL TRANSPLANT					
Drake et al. [62]	272	Various	NA	50% @ 27 mo	50% @ 25 mo
Mahindra et al. [63]	99	Various (melphalan-based 91%)	3-year: 5%	34% @ 3 yr	64% @ 3 yr
ALLOGENEIC STEM CELL TRANSPLANT					
Mahindra et al. [63]	50	MAC: 68% RIC: 32%	3-year: 41%	20% @ 3 yr	39% @ 3 yr
Morris et al. [64]	62	MAC: 73% RIC: 27%	NA	MAC: 19% @ 5 yr RIC: 11% @ 5 yr	MAC: 27% @ 5 yr RIC: 19% @ 5 yr

PFS: progression-free survival; OS: overall survival; pts: patients; mo: months; yr: year; NA: not available; STC: stem cell transplant; NRM: non-relapse mortality; MAC: myeloablative conditioning; RIC: reduced-intensity conditioning.

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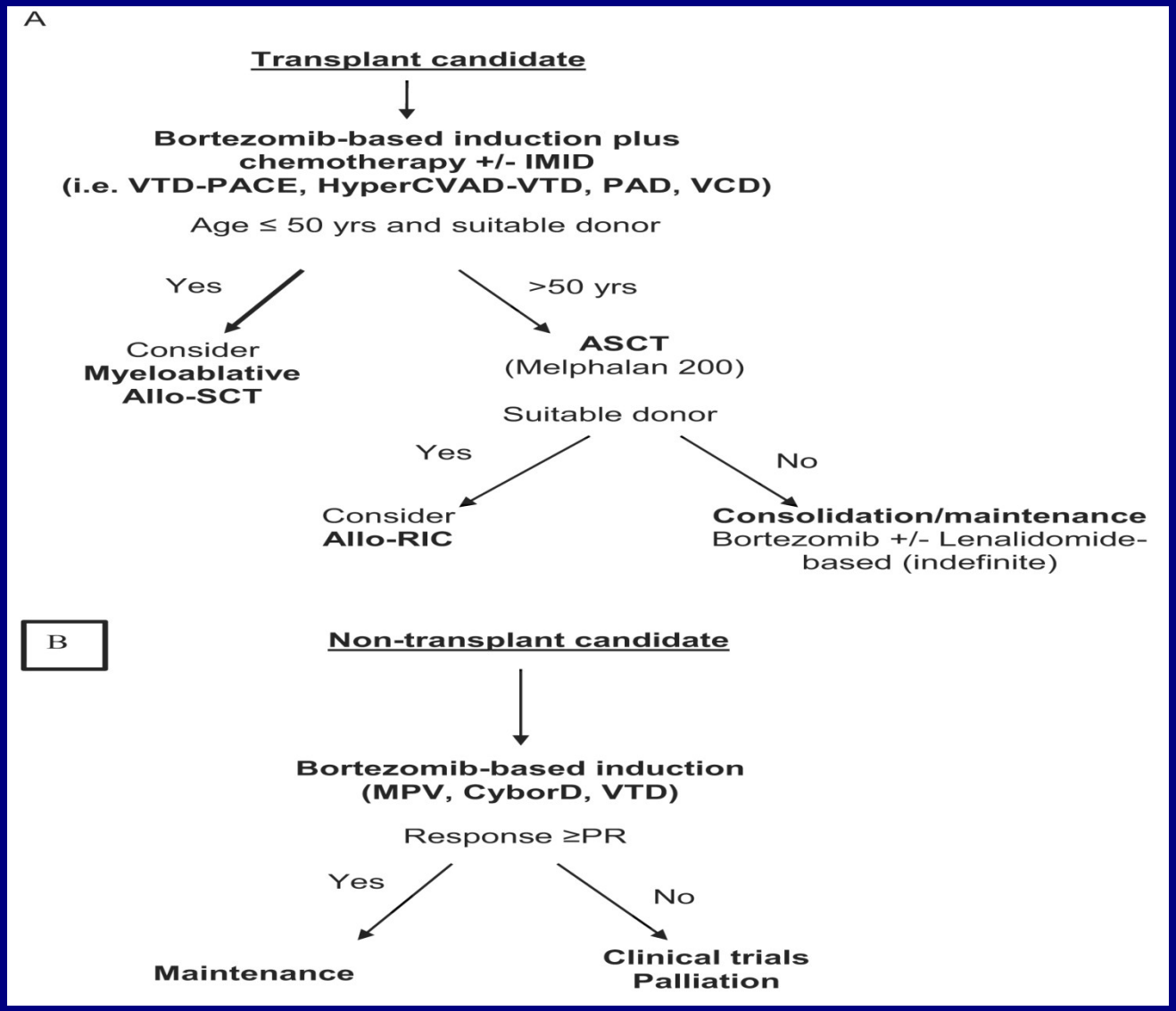
Table 1. Response rates for pPCL that underwent ASCT and maintenance.

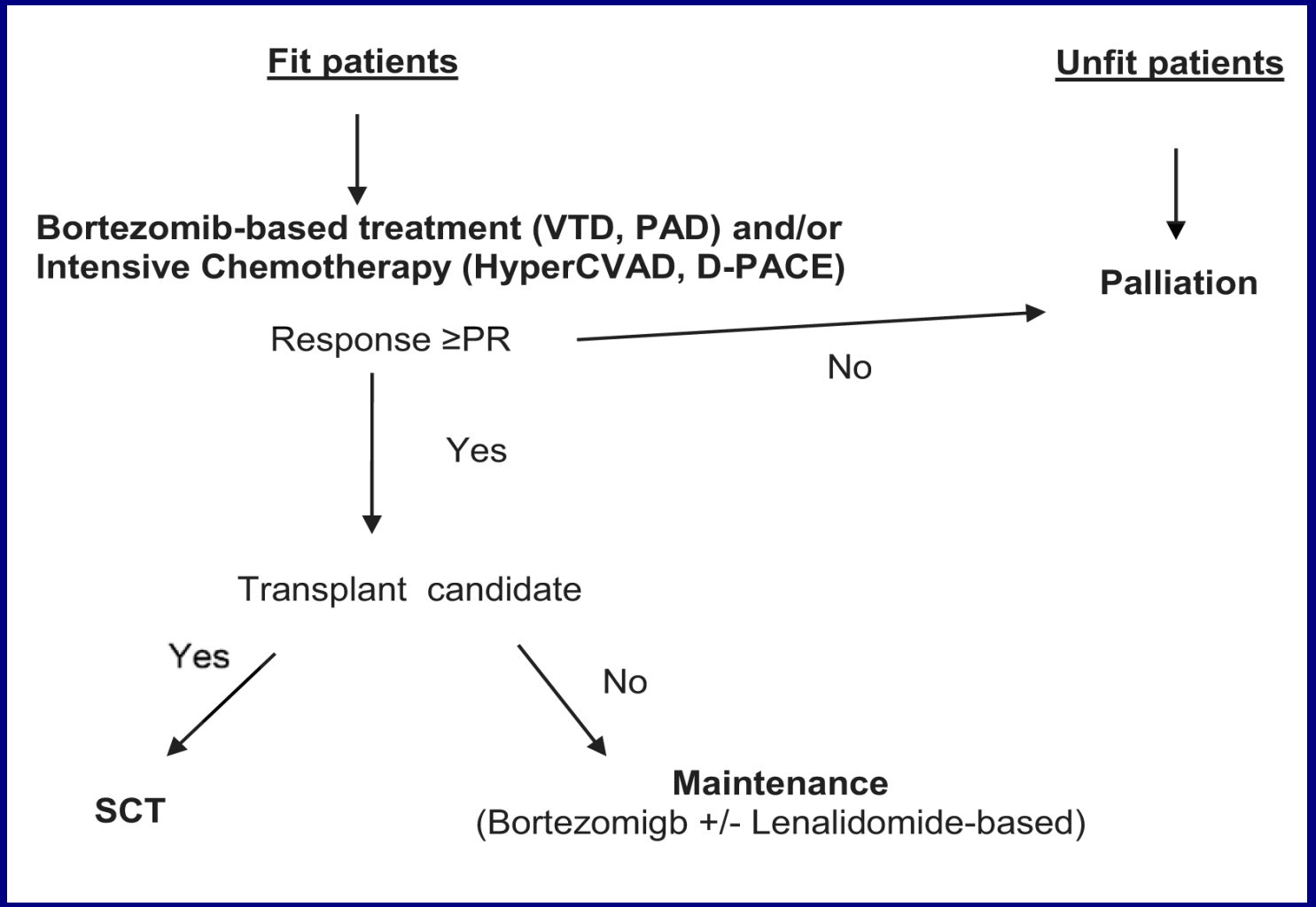
	Pre-ASCT response (n=21)		Post-ASCT response (n=20)		Best response with maintenance (n=20)	
	n	%	n	%	n	%
sCR	2	10	5	25	6	30
CR	4	20	4	20	4	20
sCR + CR	6	30	9	45	10	50
VGPR	3	15	8	40	8	40
≥VGPR	9	45	17	85	18	90
PR	10	50	1	5	1	5
≥PR	19	95	18	90	19	95
SD	0	0	0	0	1	5
PD	2	10	1	5	0	0

Treatment algorithm for primary plasma cell leukemia



Carlos Fernandez de Larrea *Leukemia. 2013 April ; 27(4): 780–791*

Treatment algorithm for secondary plasma cell leukemia or relapsed primary plasma cell leukemia



Carlos Fernandez de Larrea *Leukemia. 2013 April ; 27(4): 780–791*

DISCUSSION

- Cosa facciamo nei nostri centri ?
- Consolidamento ?
- Mantenimento ?

Documento di rete

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