

A nationwide survey of adult T-cell leukemia/lymphoma (ATL) newly diagnosed over the last decade in Japan

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Conflict of Interest disclosure by representative speakers or investigators

The authors have no financial conflicts of interest to disclose concerning the presentation.

Clinical Subtype of ATL - Shimoyama's classification by evaluating patients diagnosed during the period of 1978-87

Smouldering type

lymphocyte count $\leq 4000/\mu\text{l}$
 abnormal T lymphocyte $\geq 5\%$
 LDH $\leq 1.5\times$ normal upper limit
 \pm skin, lung involvement

Chronic type

lymphocyte count $\geq 4000/\mu\text{l}$
 abnormal T lymphocyte $\geq 3500/\mu\text{l}$
 LDH $\leq 2\times$ normal upper limit
 no Ca^{2+} ↑, CNS, bone, GI, ascites,
 or pleural effusion.

\pm LNs, skin, lung, liver/spleen

Lymphoma type

lymphadenopathy, no lymphocytosis
 abnormal T lymphocyte $\leq 1\%$

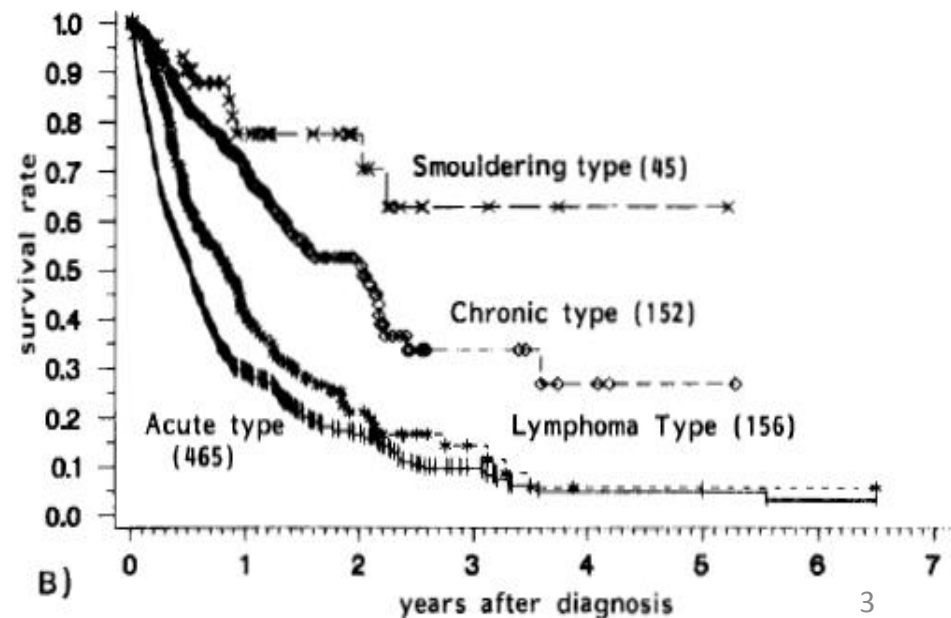
Acute type

leukemic and/or Lymphadenopathy
 excluding above 3 conditions

Shimoyama M. et al.

Br J Haematol 79: 428, 1991

	MST	2y-OS	4y-OS
acute	6.2M	16.7%	5.0%
lymphoma	10.2M	21.3%	5.7%
chronic	24.3M	52.3%	26.9%
smouldering	>60M	77.7%	62.8%



Current treatment strategy for ATL

**Diagnosis of ATL: Evidence of peripheral T-cell tumors +
Positive serum antibody against HTLV-1 (monoclonal integration of HTLV-1)**

- **Favorable chronic type**
- **Smouldering type**

**Watchful waiting until
acute transformation**

- **Acute type**
- **Lymphoma type**
- **Unfavorable chronic type***

≥ 70 y.o.

- **Reduced
chemotherapy**

≤ 69 y.o.

- **Multi-agent chemotherapy**
- **Allo-hematopoietic cell
transplantation (allo-HCT)**

*** Unfavorable chronic-type : ① BUN \uparrow ② LDH \uparrow ③ Albumin \downarrow**

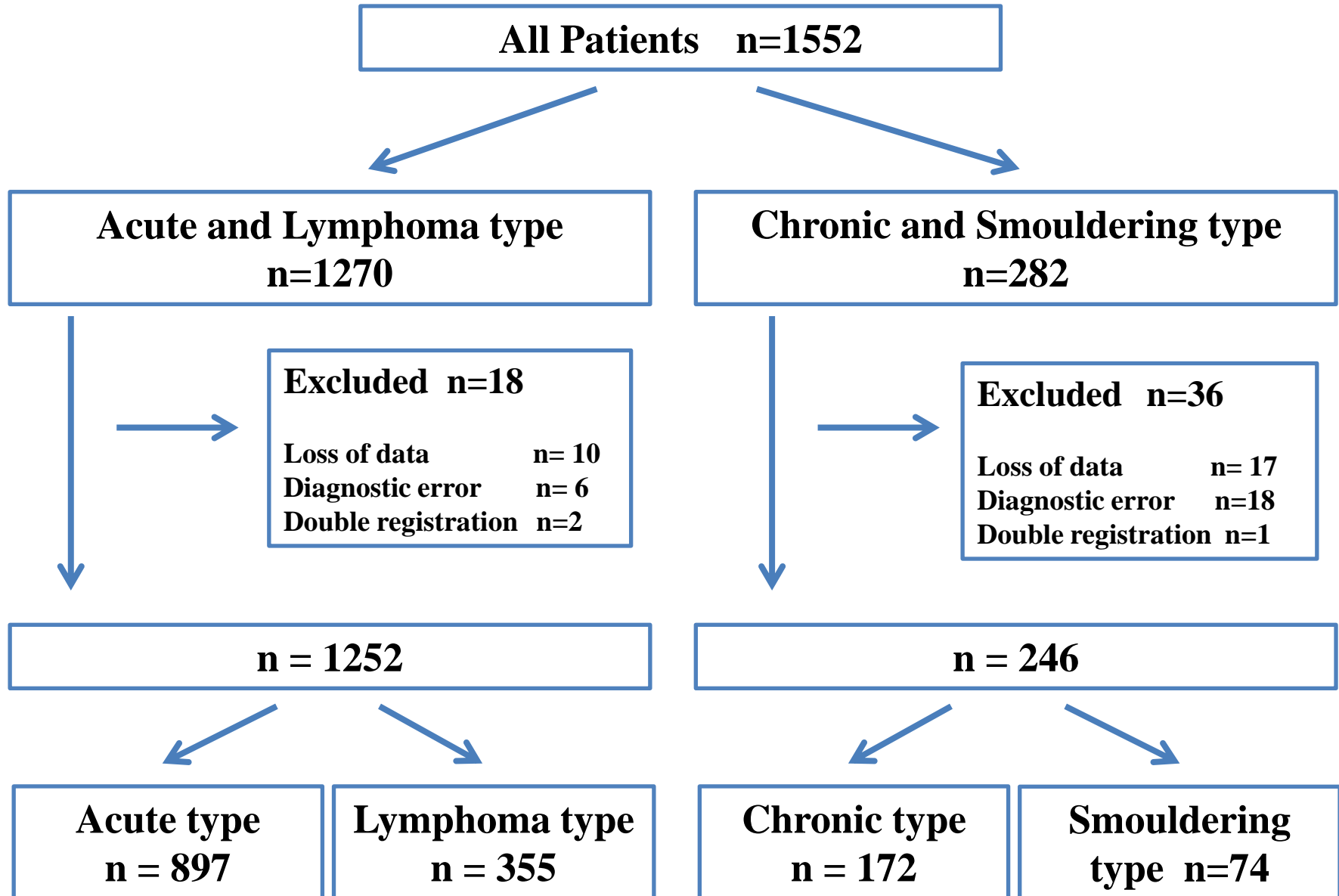
Purpose

A nationwide survey was conducted to investigate clinical features of ATL newly diagnosed during the last decade.

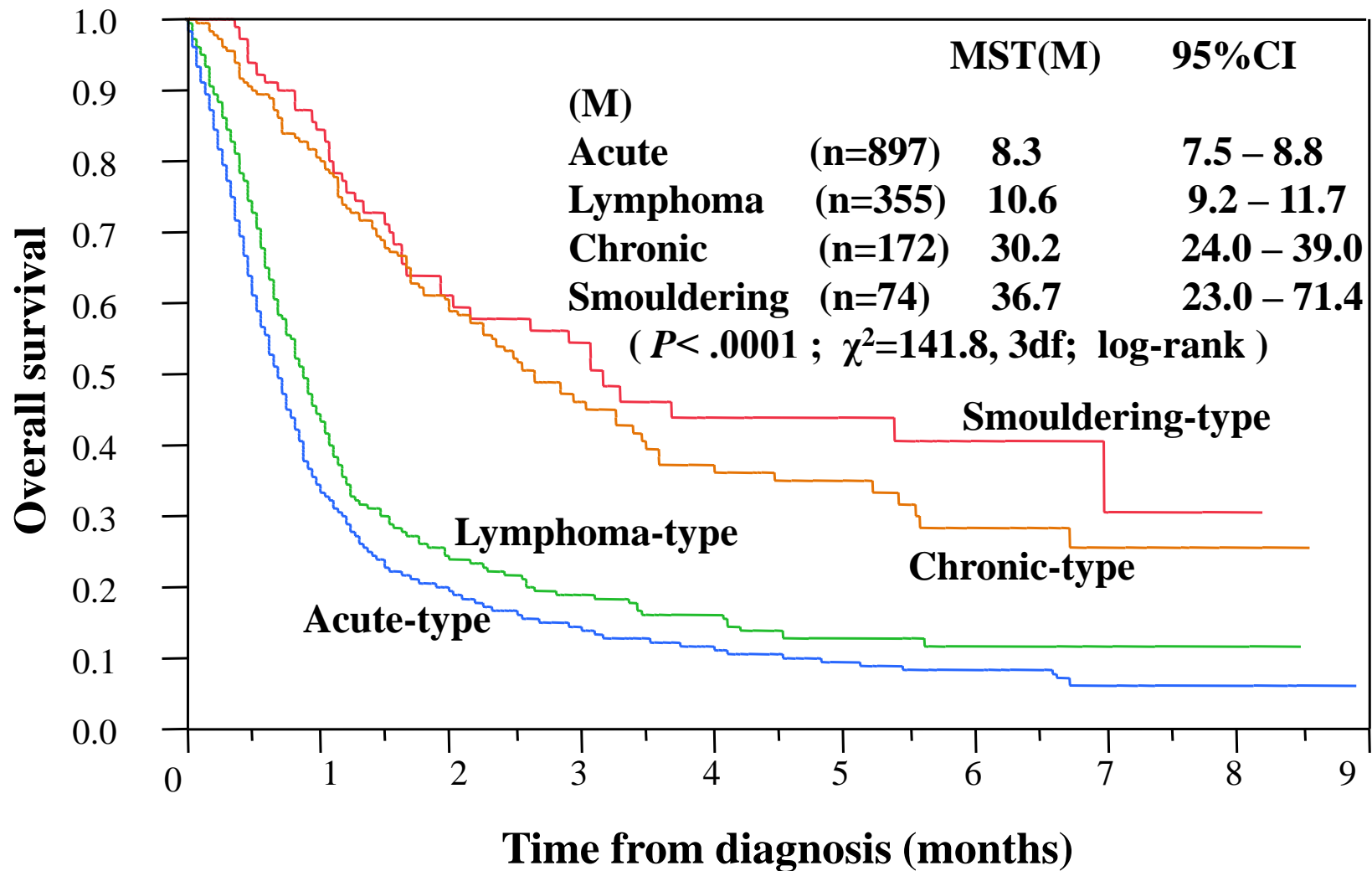
Patients and Methods

Clinical data was collected from medical records of patients who were diagnosed as ATL between 2000 and 2009 in participating institutions. Approval of this study was obtained from the Ethics Committee and Institutional Review Board of Fukuoka University where the central office is located and at each participating center based on their institutional policies.

Clinical subtypes of registered patients



Overall Survival by Clinical Subtypes



ATL

Acute and Lymphoma type

Initial Treatment for acute and lymphoma type

	n	%
Initial treatment		
Present	1188	94.9
Absent	54	4.3
Uncertain	10	0.8
total	1252	
Details of treatment		
Chemotherapy	1169	98.4
Other	19	1.6
total	1188	
Regimens of Chemotherapy		
CHOP	580	49.6
LSG15	365	31.2
ATL-GCSF	56	4.8
mEPOCH	42	3.6
Singe agent (V-16, MST-16, etc)	61	5.2
Other combination regimens	40	3.4
Uncertain	25	2.1
total	1169	

Treatment Response according to different regimens (acute and lymphoma type)

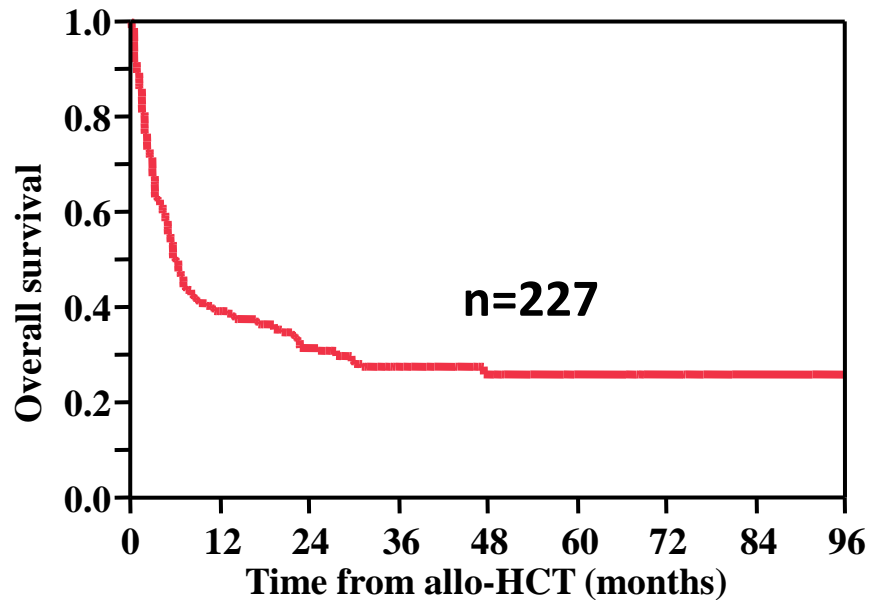
	CHOP- 14	CHOP- 21	LSG15	ATL- GCSF	mEPOCH	single	others
No of patients	140	440	365	56	43	61	40
age	62 (37-84)	68 (26-91)	61 (25-85)	56.5 (25-70)	59 (36-91)	74 (40-86)	71 (31-86)
No of course	4.8	3.9	3.1	2.5	3.2	—	—
Overall RR %	66	52	62	72	64	23	—

LSG15:VCR,CPM,ADR,PRD→ADR,MCNU,PRD→VDS,ETP,Carb,PRD

ATL-GCF:VCR,CPM,ADR,PRD, MCNU, ETP,VDS,Carb,MIT

EPOCH:ETP,PRD,VCR,CPM,ADR

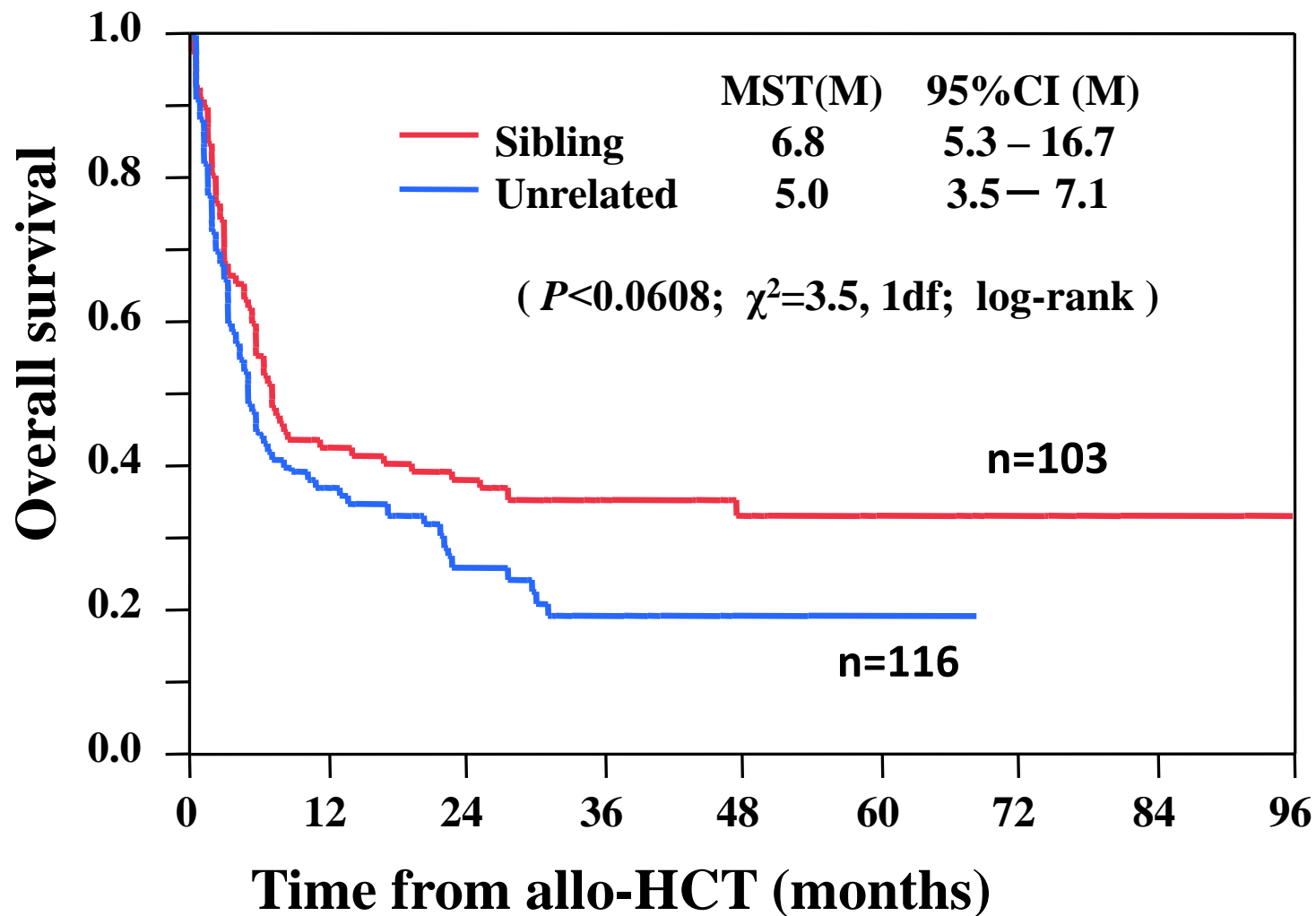
Allogeneic hematopoietic cell transplantation



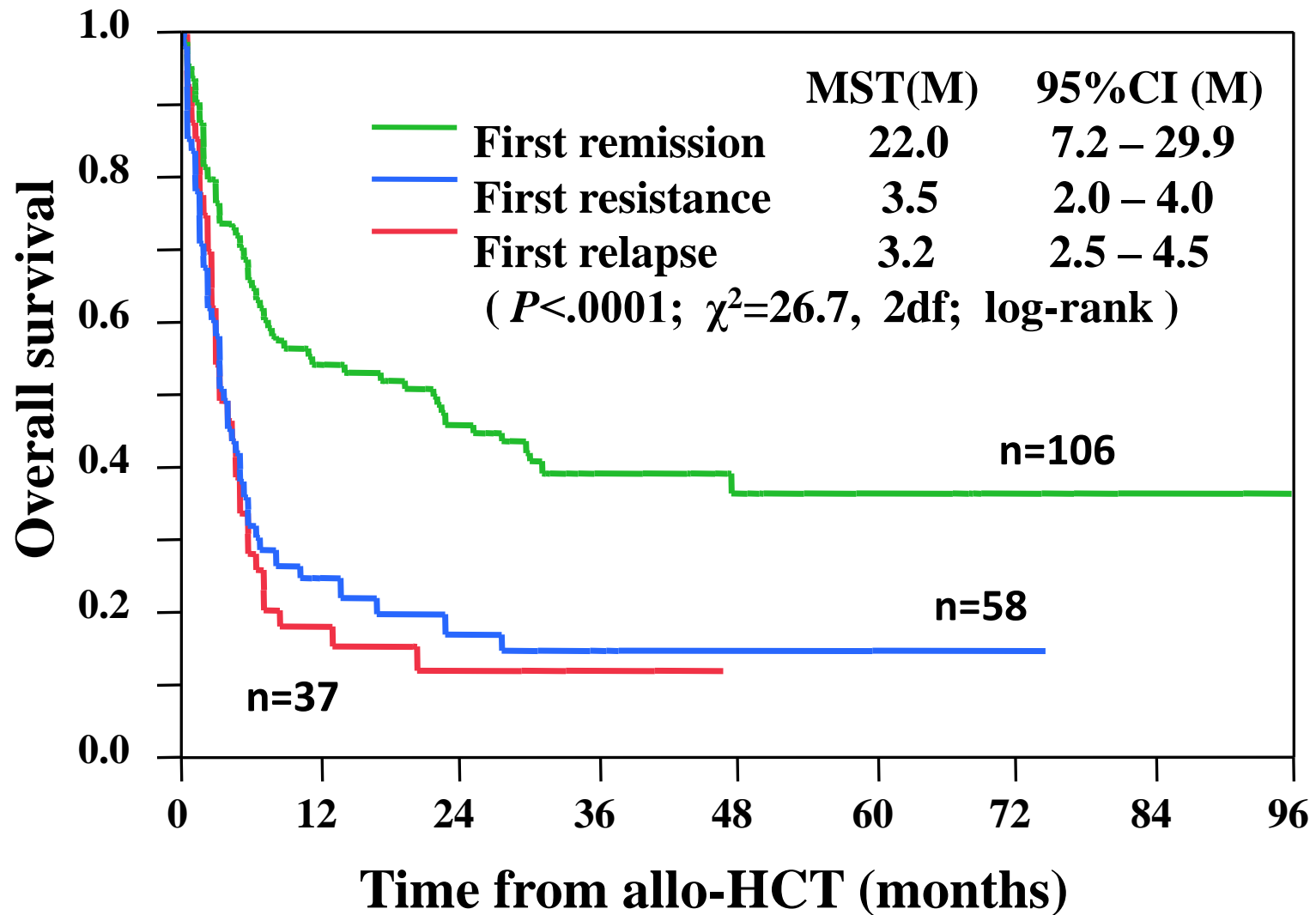
- Median age: 52 years
- 33% of acute- and lymphoma-type patients ≤ 65 years of age
- MST: 6.2 mo
- OS in 5 years: 26.0 %

	n	%
Donor		
Unrelated	116	51.1
Sibling	103	45.4
Uncertain	8	3.5
Source of stem cells		
Bone marrow	110	48.5
Peripheral blood	72	31.7
Cord blood	37	16.3
Uncertain	8	3.5
Disease status		
First remission	106	46.7
First resistance	58	25.6
First relapse	37	16.3
Others	18	7.9
Uncertain	8	3.5

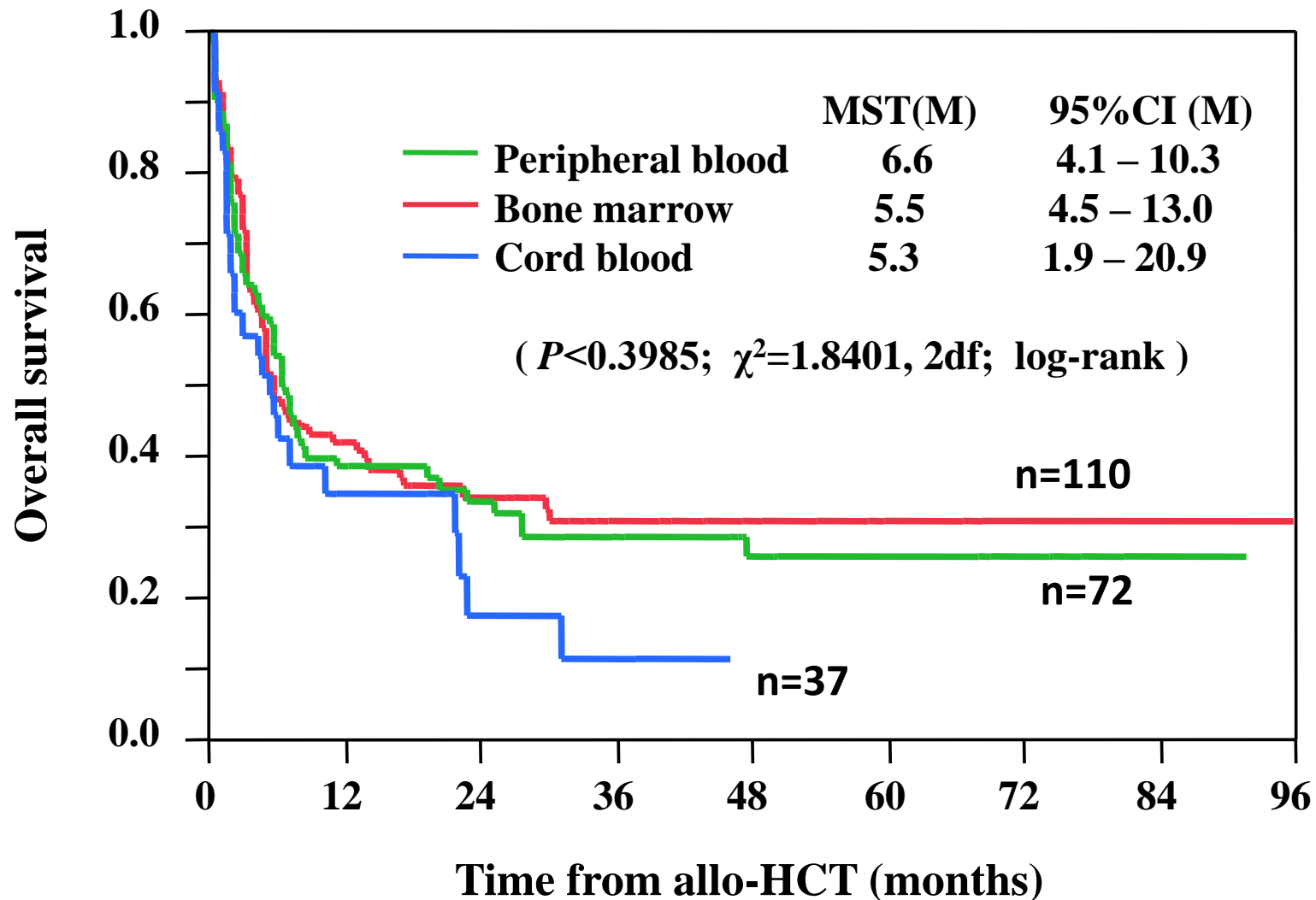
Survival according to donors



Survival according to disease status on HST



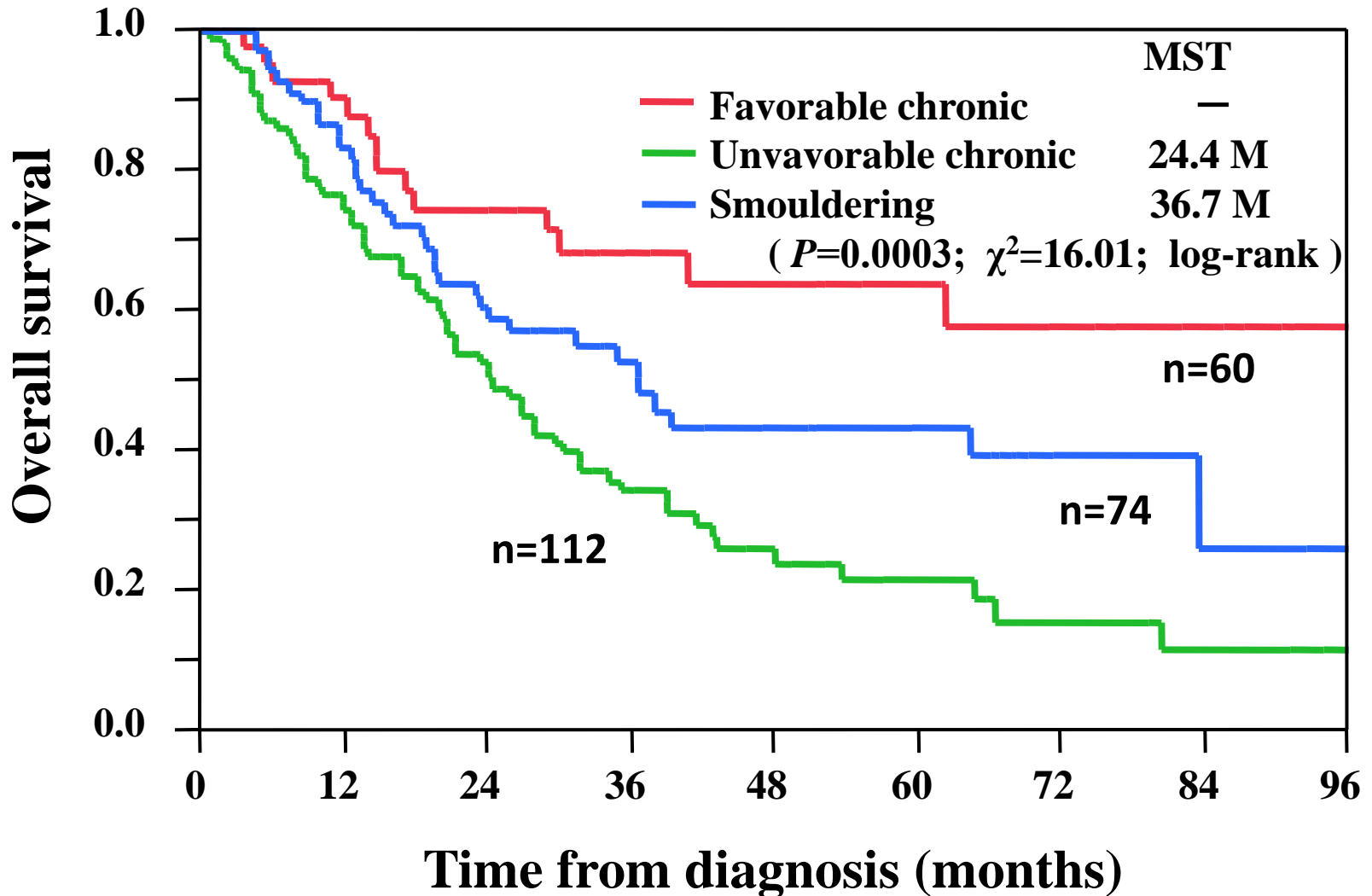
Survival according to stem cell sources



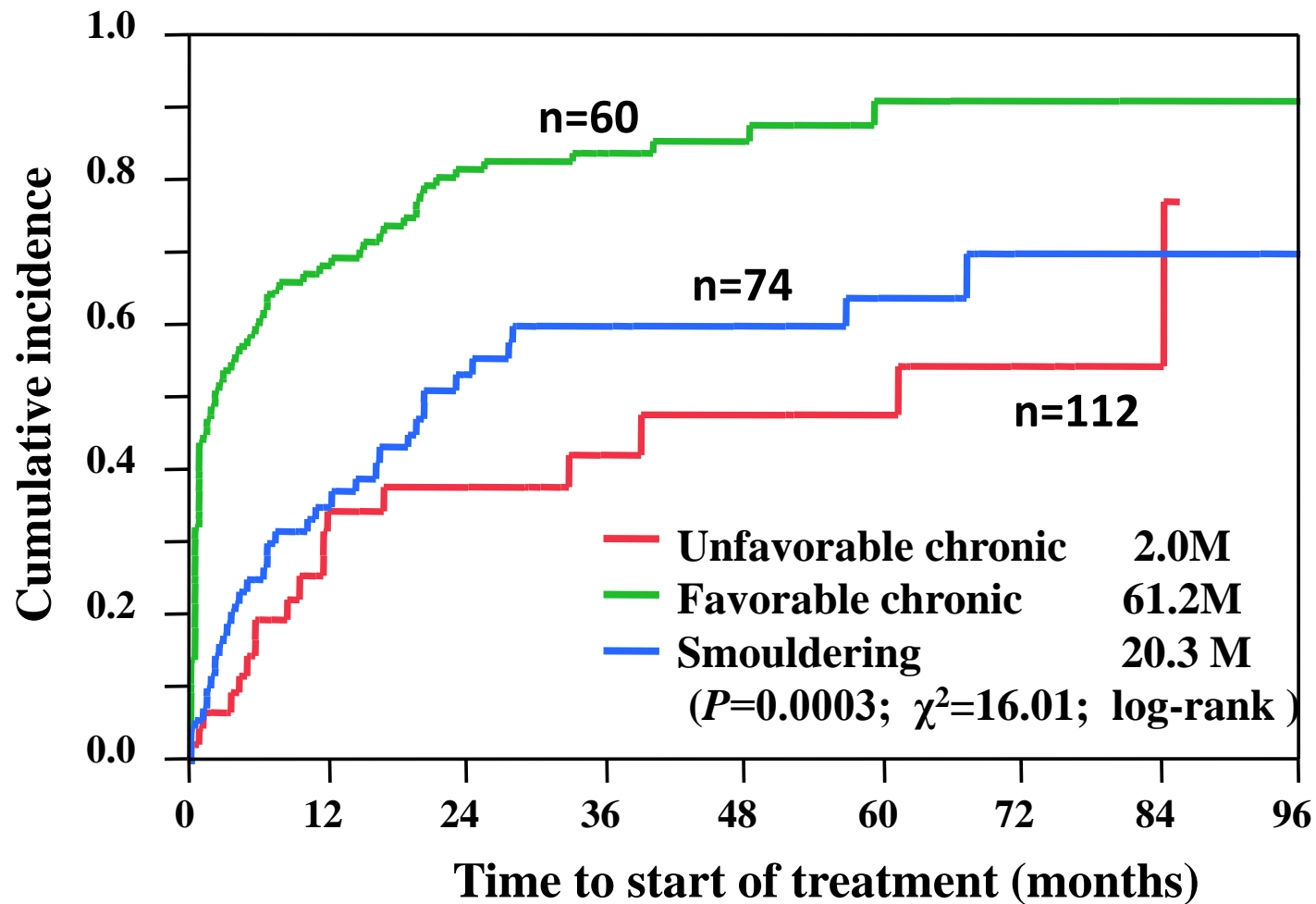
ATL

Chronic and Smouldering type

Survival for Chronic and Smouldering ATL

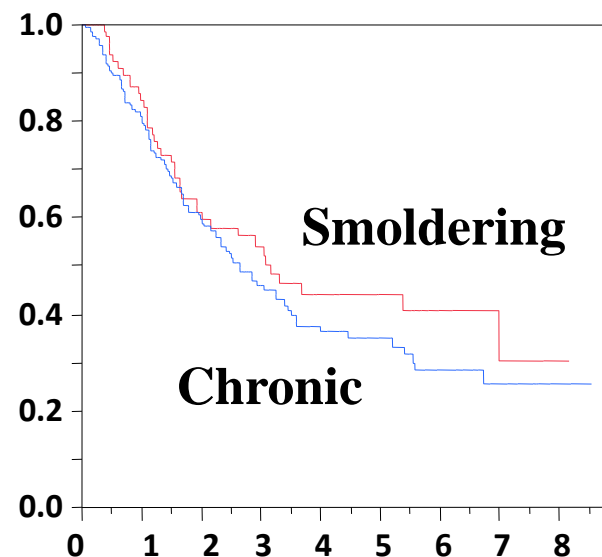
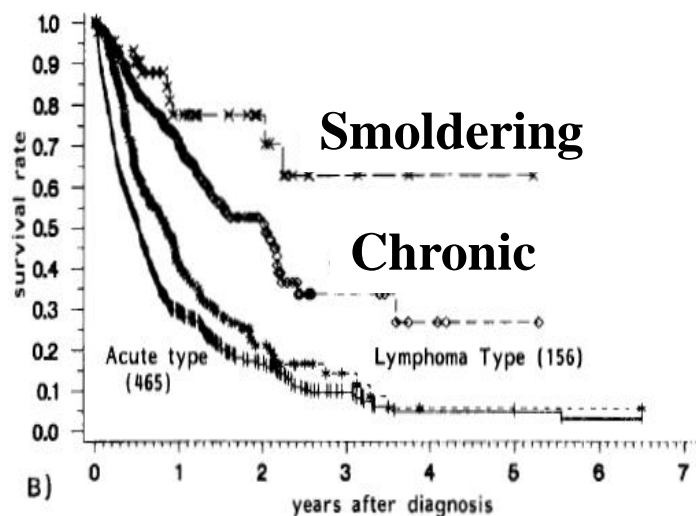


Time to start treatment for chronic and smouldering ATL



Comparison to previous report

		Shimoyama 1991 n = 197	Our data n = 246
Chronic type	median age	57.5	61.5
	MST (M)	24.3	30.3
	4-year OS (%)	26.9	35.0
	5-year OS (%)	n.a.	33.6
Smoldering type	median age	59.3	64.9
	MST (M)	not reached	36.7
	4-year OS (%)	62.8	43.2
	5-year OS (%)	n.a.	43.2



Comparison to previous report for smouldering ATL

	Shimoyama 1991	Our data
	n = 45	n = 74
Median age (year)	59.3	64.9
PS 2-4 (%)	22.7	8.2
Skin lesion, presence (%)	48.9	42.4
Lung lesion, presence (%)	15.6	3.5
No. of TIL ≥ 4 (%)	6.7	0
No. of EXNL ≥ 4 (%)	6.7	0

TIL: total involved lesion
EXNL: extranodal lesion

Conclusion

- **Prognosis of acute and lymphoma ATL is still poor despite recent progress in treatment modalities.**
- **Treatment outcome is not remarkably improved as compared to the former survey.**
- **Prognosis of smoldering type is not good as expected from the previous survey.**
- **25% of patients who underwent transplantation experienced a long survival.**