

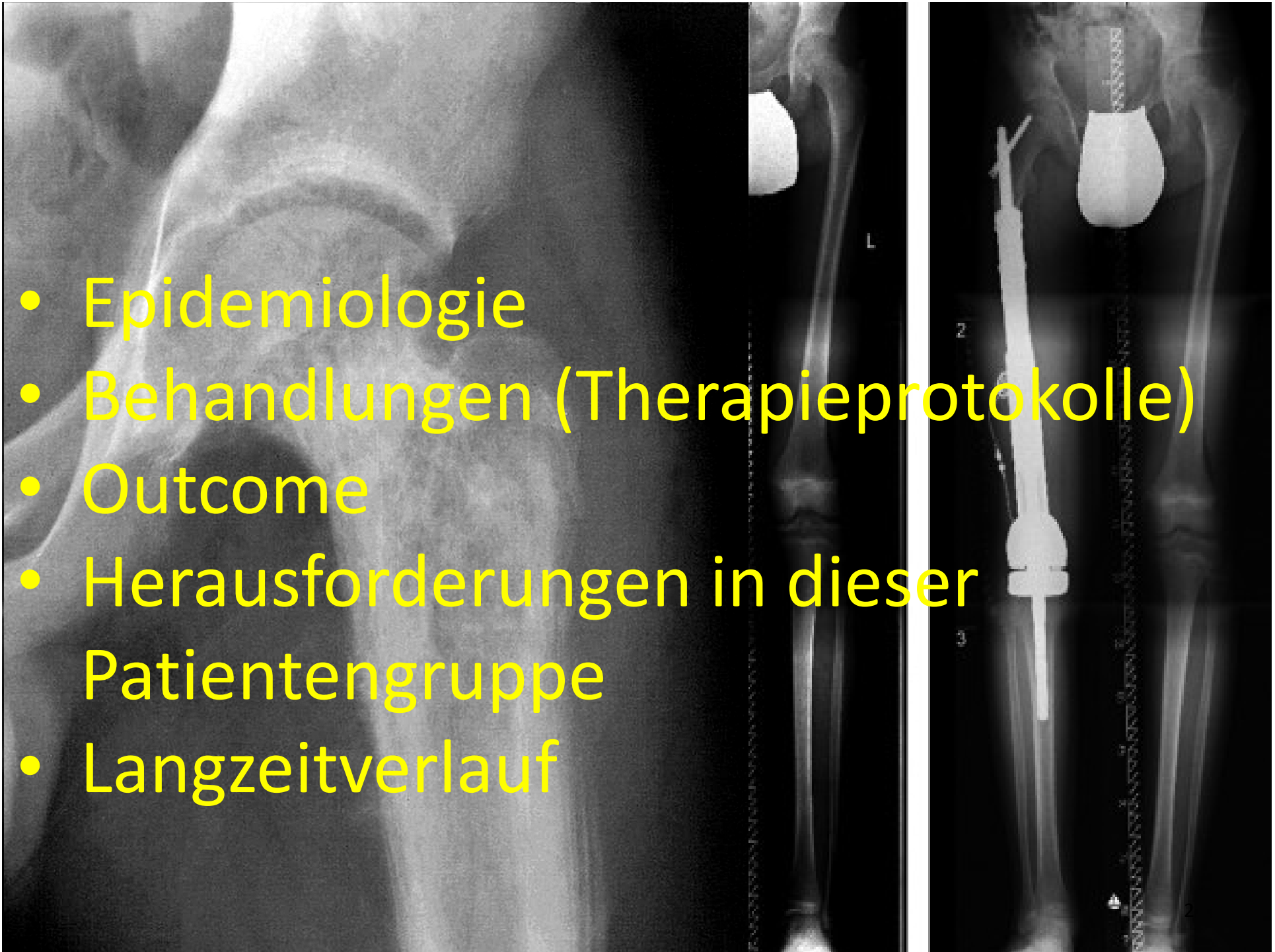


# Sarkome bei Kindern und Jugendlichen

## Sicht des pädiatrischen Onkologen

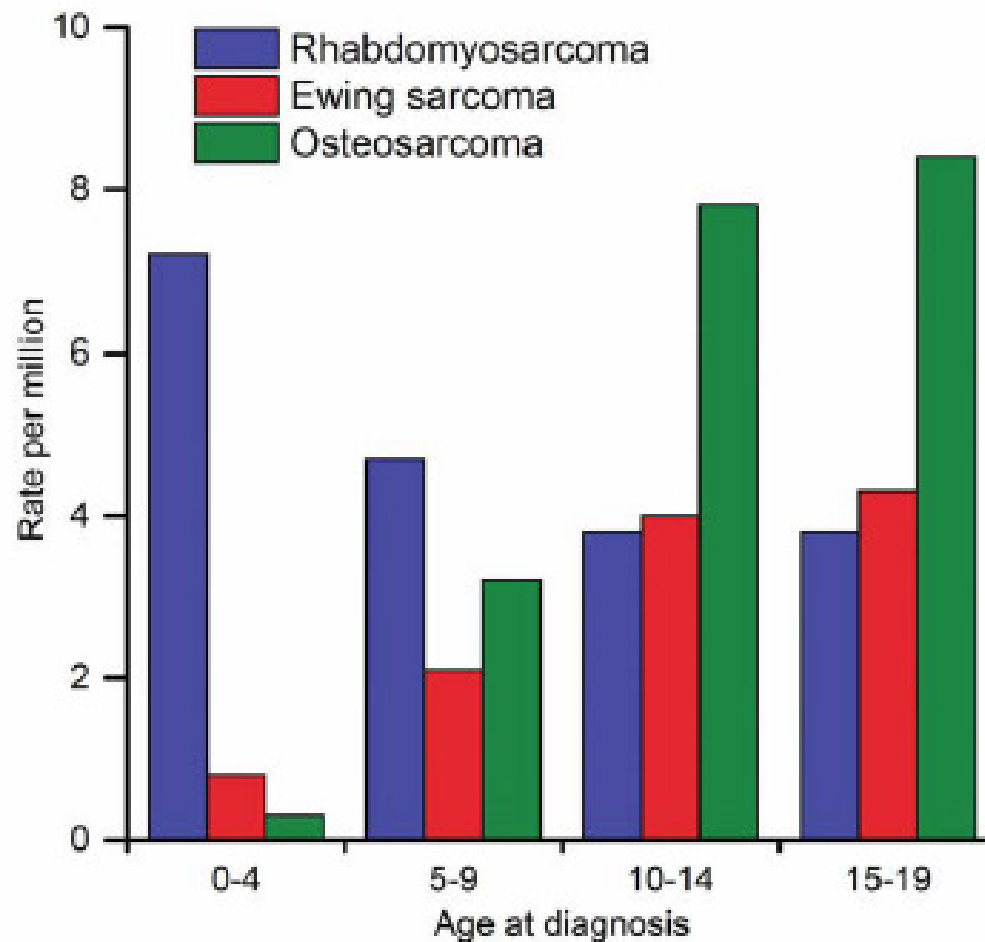


- Epidemiologie
- Behandlungen (Therapieprotokolle)
- Outcome
- Herausforderungen in dieser Patientengruppe
- Langzeitverlauf



# SEER Inzidenzraten 2011-2014

Cancer Type	Age: 0 – 14 years	Age: 0 – 19 years
all malignancies	150.6/10 <sup>6</sup> (100%)	166.4/10 <sup>6</sup> (100%)
all malign. bone tumors	6.6 (4.4%)	8.9 (5.3%)
Osteosarcoma	4.0 (2.7%)	5.2 (3.1%)
Chondrosarcoma	--	0.4 (0.2%)
Ewing and related S.	2.1 (1.4%)	2.6 (1.6%)
other malign. bone tu.	0.3 (0.2%)	0.5 (0.3%)
all STS	10.7 (7.1%)	12.0 (7.2%)
Rhabdomyosarcoma	5.2 (3.5%)	4.8 (2.9%)
Fibrosarcoma and rel.S.	1.3 (0.9%)	1.5 (0.9%)
other specified STS	3.4 (2.3%)	4.7 (2.8%)
Unspecified STS	0.9 (0.6%)	1.0 (1.0%)



**FIGURE 8. Age-Specific Incidence Rates for Bone and Soft Tissue Sarcomas, United States, 2006 to 2010.**

Source: North American Association of Central Cancer Registries. Data are included from all US states and the District of Columbia except Arkansas, Minnesota, Nevada, Ohio, and Virginia.

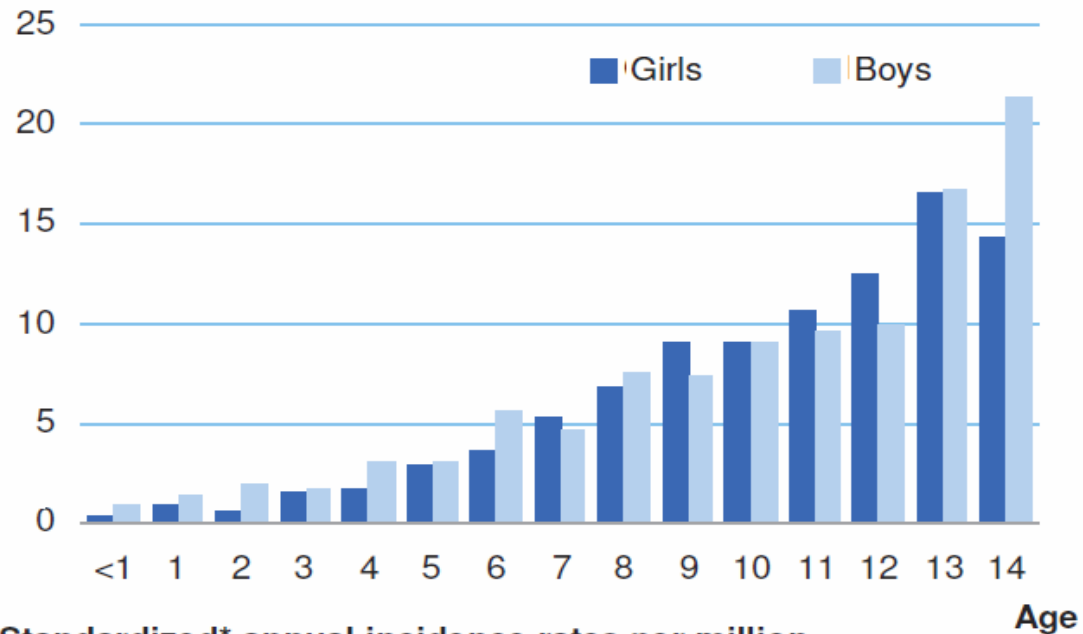
CA Cancer J Clin 2014;64:83-103. VC 2014 American Cancer Society.

# Maligne Knochtumore <15 jährig

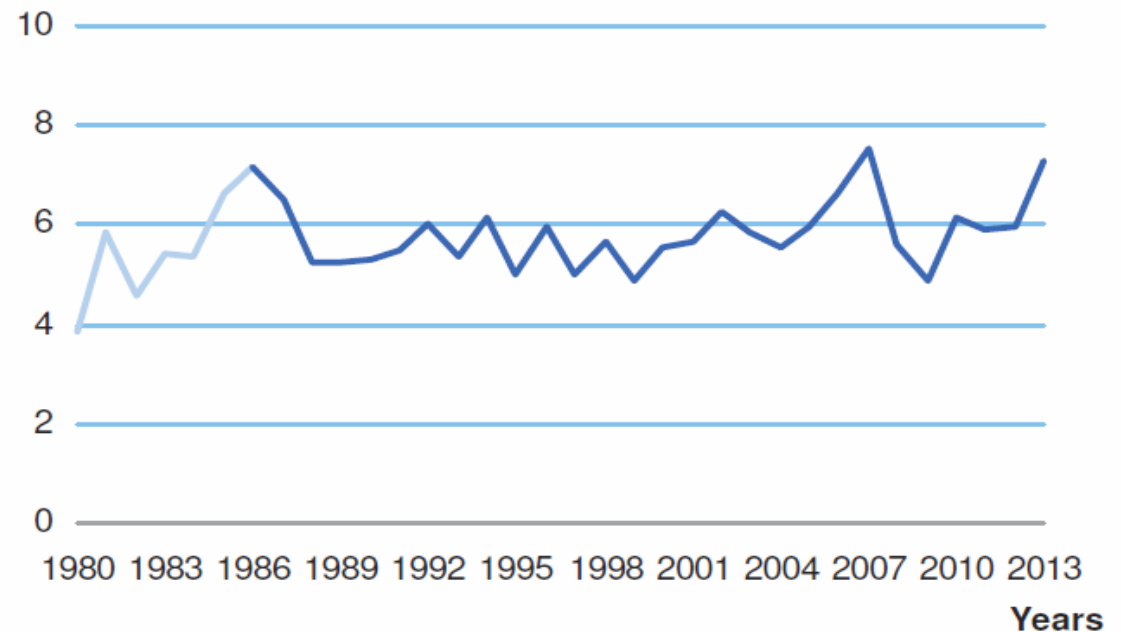
- ✓ Osteosarkome
- ✓ Chondrosarkome
- ✓ Ewing Tumore und verwandte Knochensarkome
- ✓ Andere spezifizierte maligne Knochtumore
- ✓ Unspezifizierte maligne Knochtumore

Deutsches Kinderkrebsregister, JB 2013/14

Age- and sex-specific incidence rates per million  
Germany 2004-2013

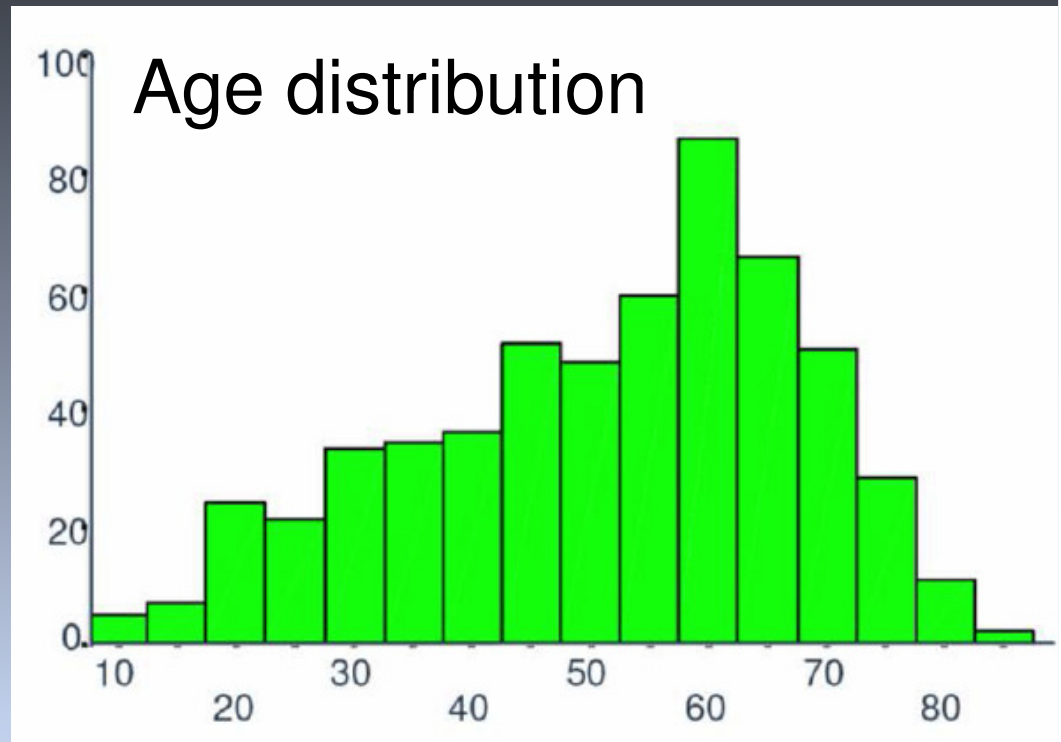


Standardized\* annual incidence rates per million  
Germany 1980-2013



- ✓ STS  $\approx$  1% aller Malignitäten
- ✓ Bei Patienten <15 jähig: 0.5-1/100'000
- ✓ 30% >60 jähig
- ✓ >50 histologische Subtypen
- ✓ Kinder- und Jugendliche: chromosomale Translocationen, Fusionsgene
- ✓ Ältere Patienten: hochkomplexe aberrante Genome

## Weichteilsarkom (STS)

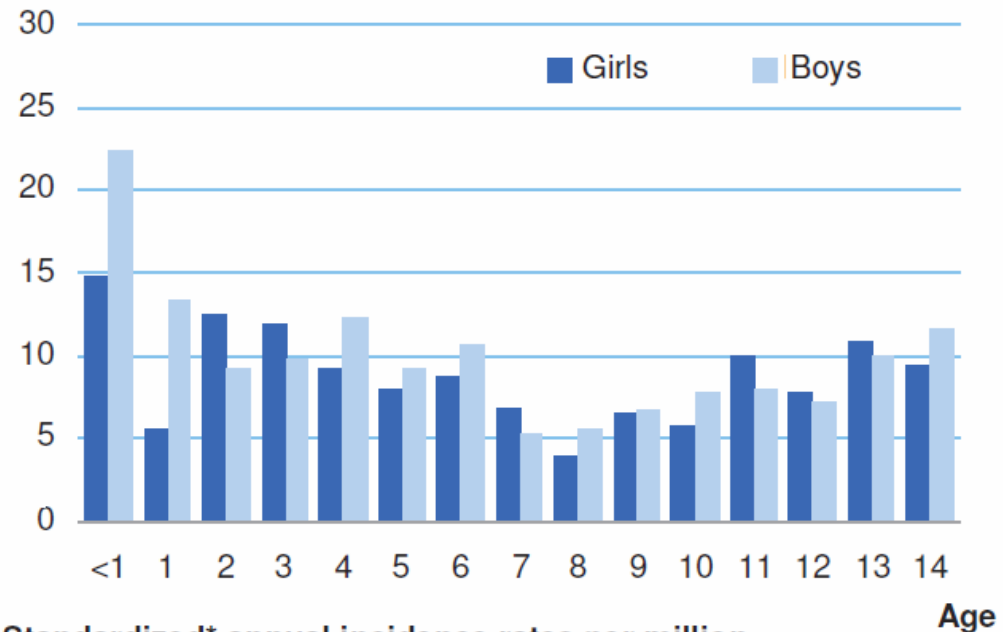


## Weichteil und andere extraossäre Sarkome < 15 jährig

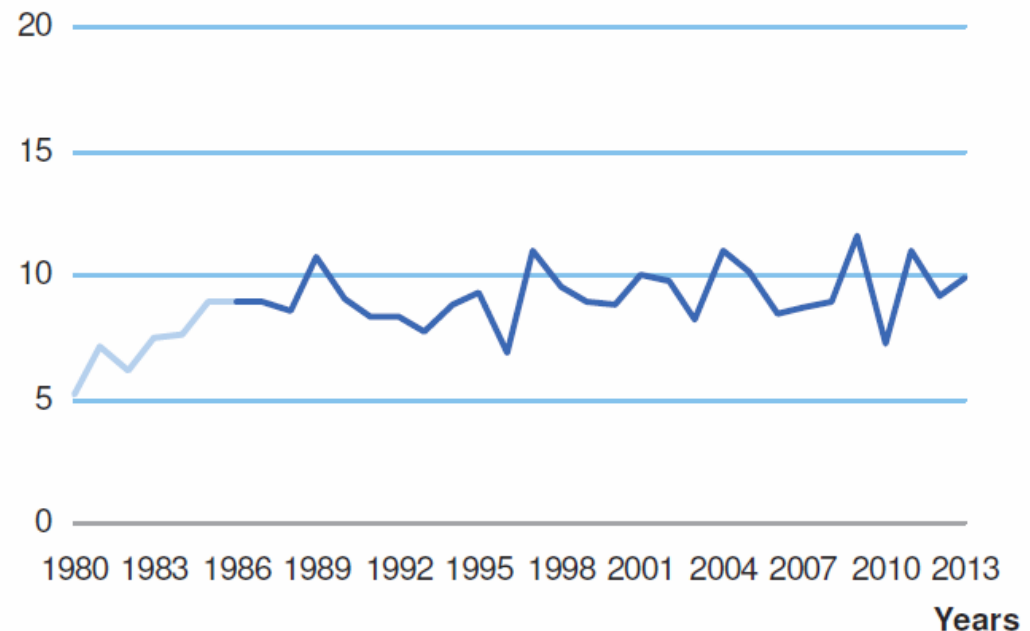
- ✓ Rhabdomyosarkome
- ✓ Fibrosarkome, peripherer Nervenscheidentumore und andere fibröse Neoplasien
- ✓ Kaposi Sarkome
- ✓ Andere spezifizierte maligne Weichteilsarkome
- ✓ Unspezifizierte maligne Weichteilsarkome

Deutsches Kinderkrebsregister, JB 2013/14

Age- and sex-specific incidence rates per million  
Germany 2004-2013



Standardized\* annual incidence rates per million  
Germany 1980-2013





- Epidemiologie
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- Outcome
- Herausforderungen in dieser Patientengruppe
- Langzeitverlauf





Alter: 0 bis < 40 Jahre  
2260 Patienten  
eingeschlossen  
>50% randomisiert



**CureSearch**  
Children's Oncology Group



**E**UROPEAN  
**O**STEOSARCOMA  
**I**NTERGROUP



GESELLSCHAFT FÜR  
PEDIATRISCHE ONKOLOGIE  
UND HÄMATOLOGIE **GPOH**



**NCRI**  
National  
Cancer  
Research  
Institute



Scandinavian Sarcoma Group  
1979

THE BONE AND JOINT DECADE 2000-2010  
**Participating  
Organization**

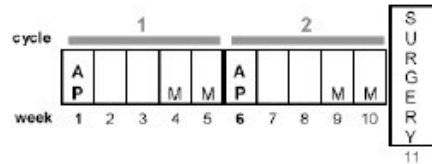
## EURAMOS 1

ISRCTN67613327  
EudraCT no. 2004-000242-20

A randomized trial of the  
European and American Osteosarcoma Study Group  
to optimize treatment strategies for resectable  
osteosarcoma based on histological response to  
pre-operative chemotherapy

Clinical trial protocol  
Version: 1.2  
Date: 30 March 2007

A = Doxorubicin 75mg/m<sup>2</sup>/course  
 P = Cisplatin 120mg/m<sup>2</sup>/course  
 M = Methotrexate 12g/m<sup>2</sup>/course  
 E = Etoposide 300mg/m<sup>2</sup>/course  
 I = Ifosfamide 14g/m<sup>2</sup>/course  
 i = Ifosfamide 9g/m<sup>2</sup>/course  
 Ifn = Interferon- $\alpha$  0.5-1.0 $\mu$ g/kg weekly

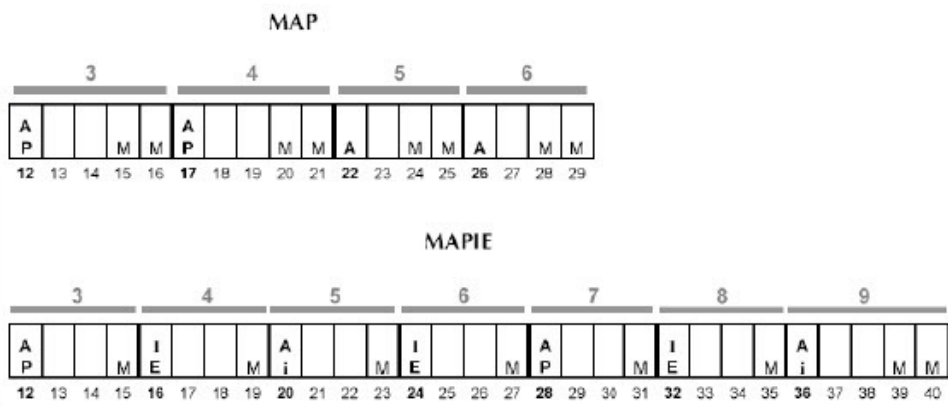
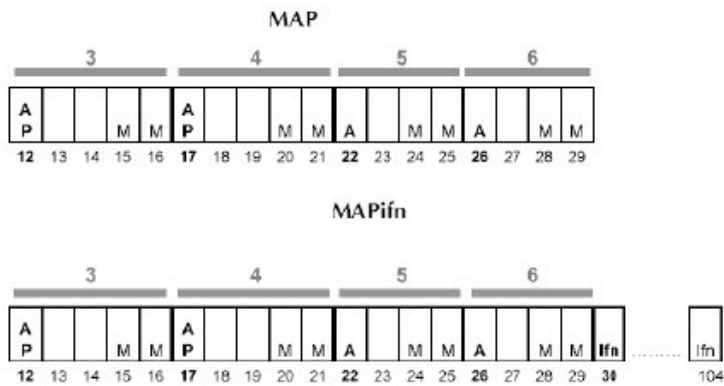


GOOD RESPONSE

**RANDOMISE**

POOR RESPONSE

Evaluation of histological response



Note: Surgery for metastases should take place between weeks 11-20. See section 9.2.2.5



In collaboration with



## EWING 2008

EudraCT number: 2008-003658-13

Trial Code: EWING2008

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[Uta.Dirksen@ukmuenster.de](mailto:Uta.Dirksen@ukmuenster.de)

Planned period of study: 2009-2018

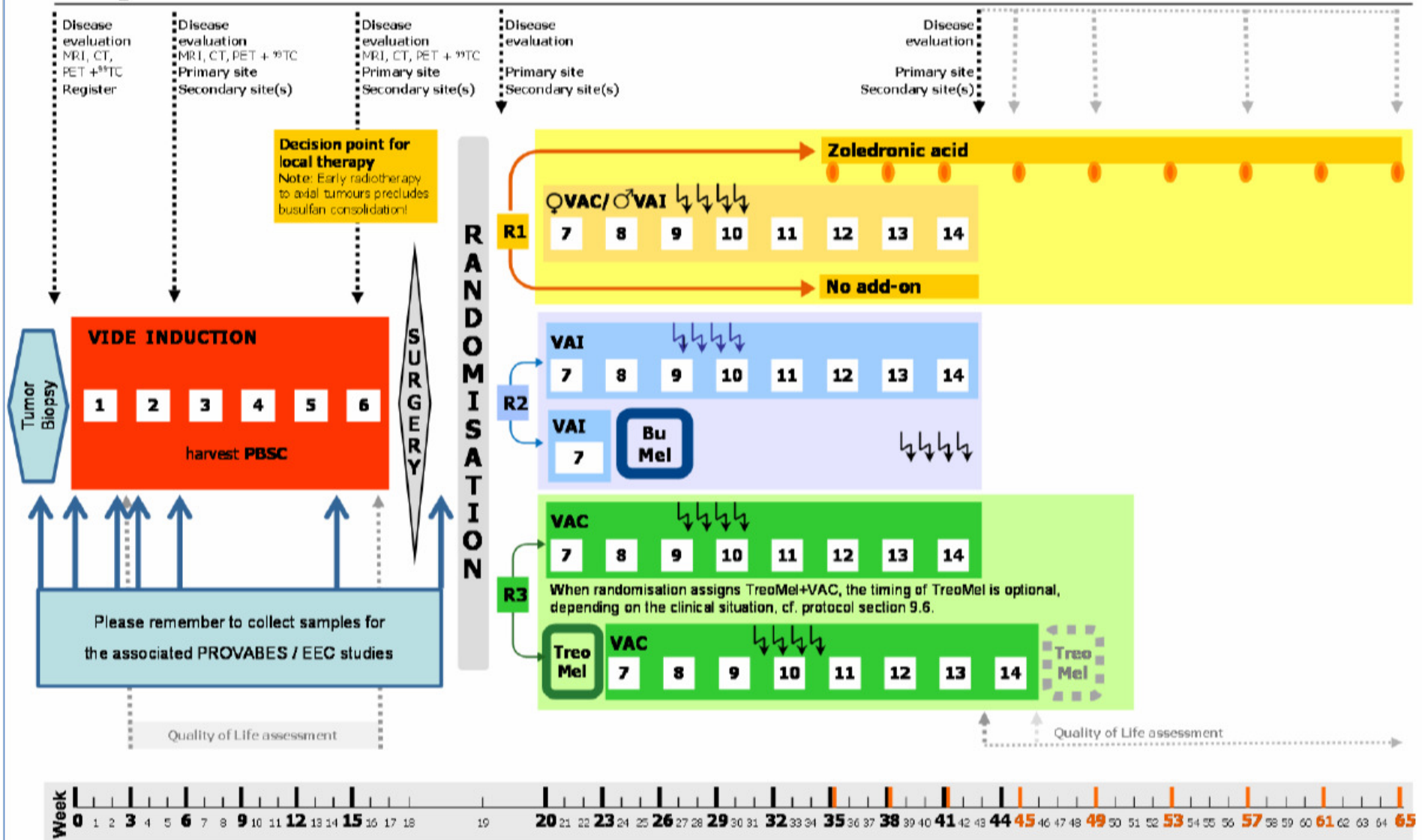
Version: 1.9

Date of version: 1 Oct 2014

Alter: >4 bis <50 Jahre  
Ziel: n = 1383  
Patienten



# Ewing 2008 Treatment Schema



## Cooperative Weichteilsarkom Studiengruppe CWS der GPOH



# CWS-2007-HR

A randomised phase-III trial of the  
Cooperative Weichteilsarkom Studiengruppe  
for localised high-risk Rhabdomyosarcoma and  
localised Rhabdomyosarcoma-like Soft Tissue Sarcoma  
in children, adolescents, and young adults

*funded by the Deutsche Kinderkrebsstiftung (German Childhood Cancer Foundation)*

*Sponsor: Universitätsklinikum Tübingen  
conducted under the auspices of the  
German Society for Paediatric Haematology and Oncology (GPOH)  
Version 1.2 from 20.02.2015*

*Clinical Trial Number (EudraCT): 2007-001478-10  
Prüfplancode des Sponsors: Z3*



Alter:  
> 6 Monate  
und  
< 21 Jahre

Cooperative Weichteilsarkom Study  
Group CWS der GPOH  
in cooperation with the  
European paediatric Soft Tissue Sarcoma Study Group  
EpSSG



## CWS-guidance

for risk adapted treatment of  
soft tissue sarcoma and soft tissue tumours  
in children, adolescents, and young adults

*Version 1.6. from 12.12.2012*



DEUTSCHE  
KREBSGESELLSCHAFT E.V.

Chair persons

Prof. Dr. med. Ewa Koscielniak  
Prof. Dr. med. Thomas Klingebiel



Cooperative Soft Tissue Sarcoma  
Study Group CWS  
Cooperative Weichteilsarkom Studiengruppe CWS der GPOH



## CWS-Register

## SoTiSaR

A Registry for soft tissue sarcoma  
and other soft tissue tumours  
in children, adolescents, and young adults

*Version 1.4 from 1.07.2009*



Chair persons

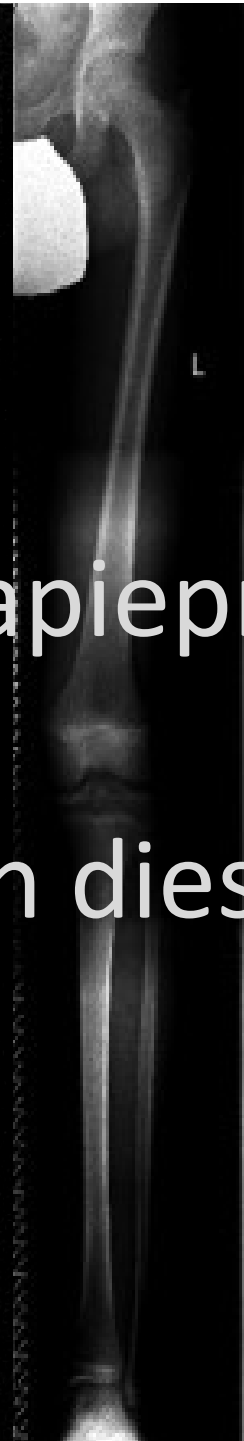
Prof. Dr. med. Ewa Koscielniak  
Prof. Dr. med. Thomas Klingebiel



### Therapy Overview CWS-Guidance at a glance

RMS-Low	Subgroup A no RTX	VA		VA		VA		VA				
	Subgroup B no RTX	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	Local Treatment	VA	VA	VA	VA	Local Control Assessment	
RMS-Standard	Subgroup C +/- RTX	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA		(I <sup>2</sup> )VA	(I <sup>2</sup> )VA	(I <sup>2</sup> )VA	(I <sup>2</sup> )VA		(I <sup>2</sup> )VA
	Subgroup D RTX	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> V(A)		I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA		I <sup>2</sup> VA
RMS-High	Subgroup E+F+G	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA		I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA	I <sup>2</sup> VA		I <sup>2</sup> VA
		VAIA III			VAIA III							
RMS-Very High	Subgroup H	I <sup>2</sup> VAd	I <sup>2</sup> VA	I <sup>2</sup> VAd	I <sup>2</sup> Vad	Local Treatment	I <sup>2</sup> VA	I <sup>2</sup> VAd	I <sup>2</sup> VA	I <sup>2</sup> VA	Local Control Assessment	
Other "RMS-like"		I <sup>2</sup> VAd	I <sup>2</sup> VA	I <sup>2</sup> VAd	I <sup>2</sup> VAd		I <sup>2</sup> VA	I <sup>2</sup> VAd	(I <sup>2</sup> )VA	(I <sup>2</sup> )VA		(I <sup>2</sup> )VA
"Non-RMS-like"		I <sup>2</sup> VAd	I <sup>2</sup> VA	I <sup>2</sup> VAd	I <sup>2</sup> VAd		I <sup>2</sup> VA	I <sup>2</sup> VAd	I <sup>2</sup> VA	I <sup>2</sup> VA		I <sup>2</sup> VA
		CEVAIE			CEVAIE			CEVAIE				
Metastatic STS		I <sup>3</sup> VA	CEV	I <sup>3</sup> VE	I <sup>3</sup> VA		CEV	I <sup>3</sup> VE	I <sup>3</sup> VA	CEV	I <sup>3</sup> VE	8x O-TIE
	Time [weeks]	1	4	7	10		13	16	19	22	25	28-52

- Epidemiologie
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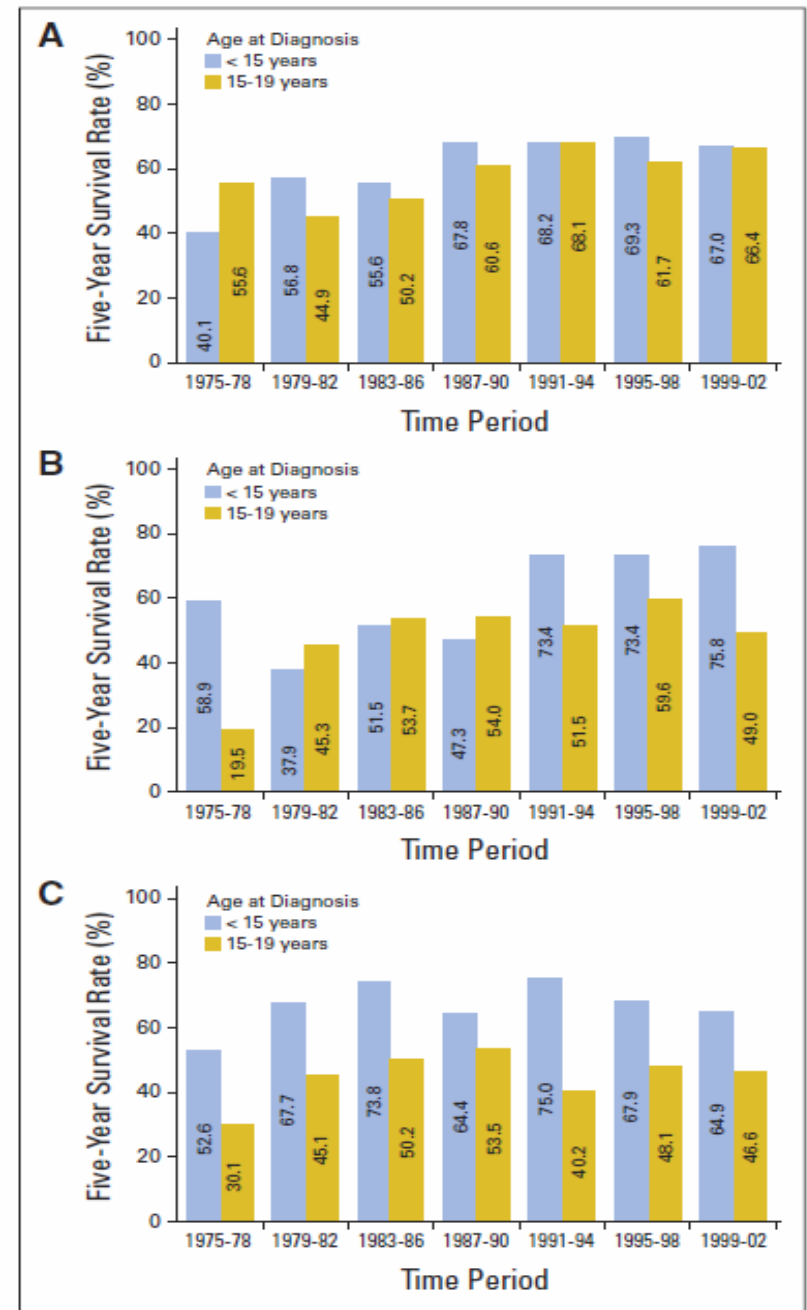


Osteosarkom

Ewing Sarkom

Rhabdomyosarkom

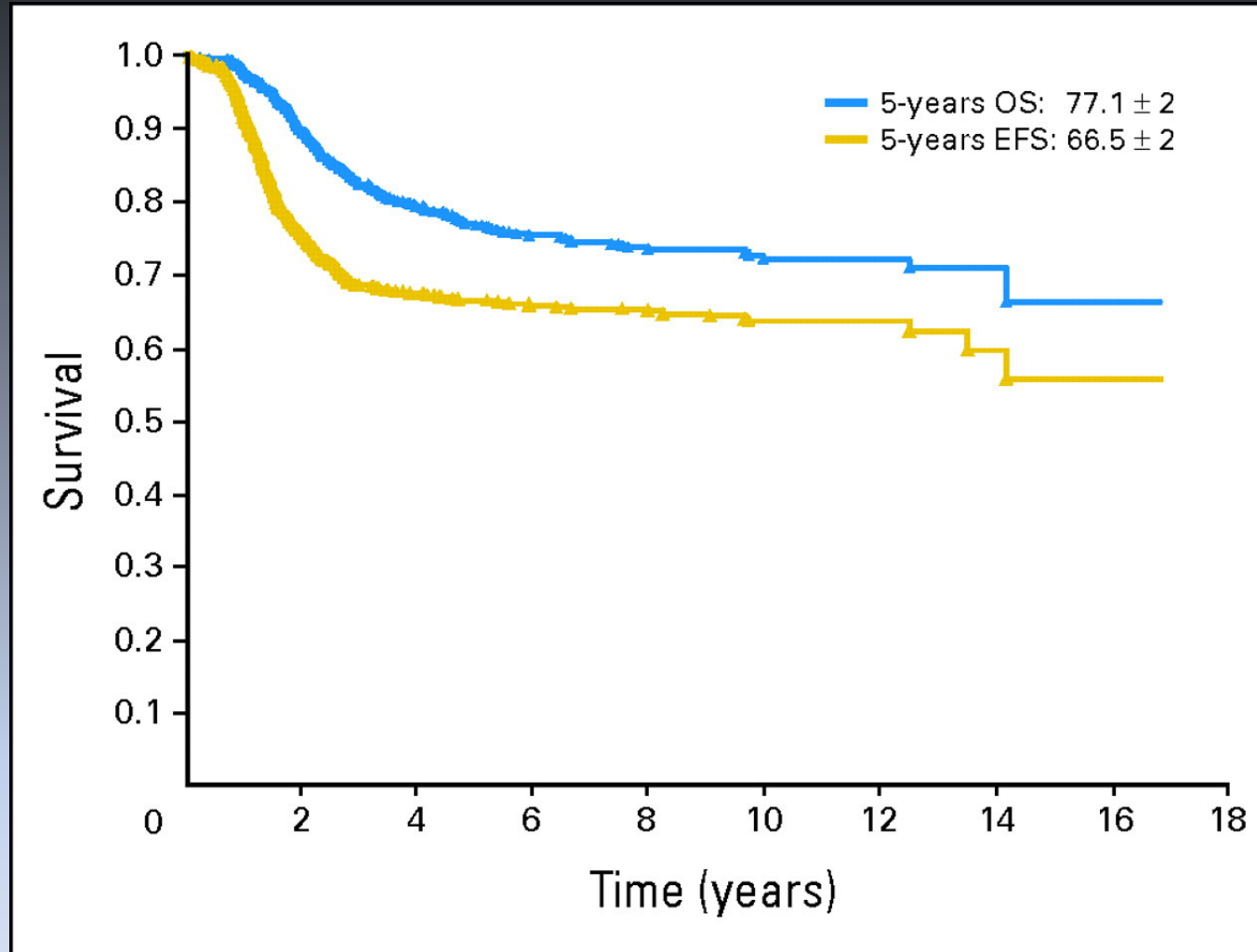
J Clin Oncol 28:2625-2634 (2010)



**Fig 7.** Five-year survival rates for (A) osteosarcoma, (B) Ewing sarcoma, and (C) rhabdomyosarcoma among children by age group and time period of diagnosis from 1975 through 2002, with follow-up of vital status through 2006, according to data from Surveillance, Epidemiology, and End Results 9 (SEER 9) registries.

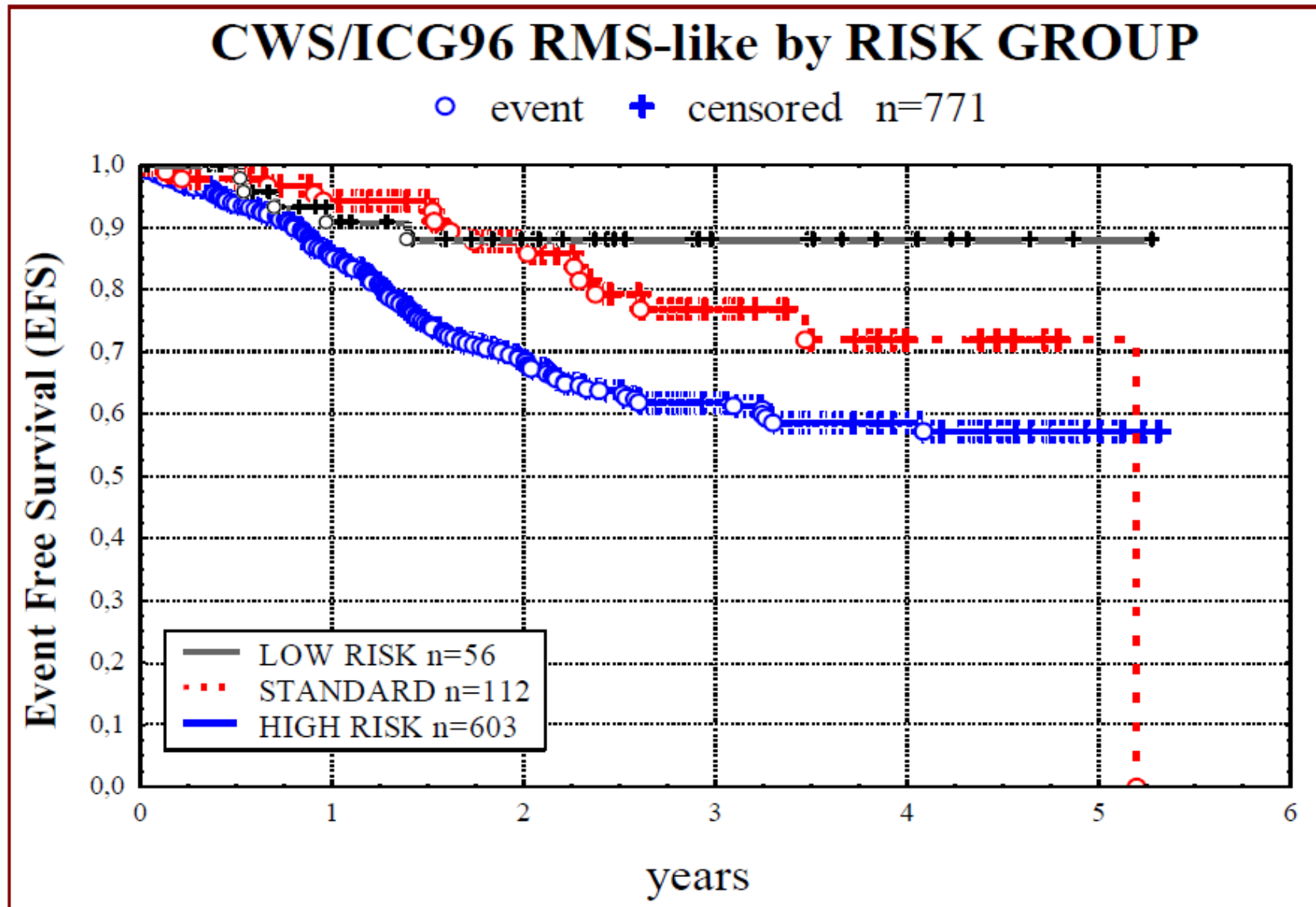


# 1578 Patienten < 21 jährig mit lokalisiertem RMS (CWS-81, CWS-86, CWS-91, CWS-96)

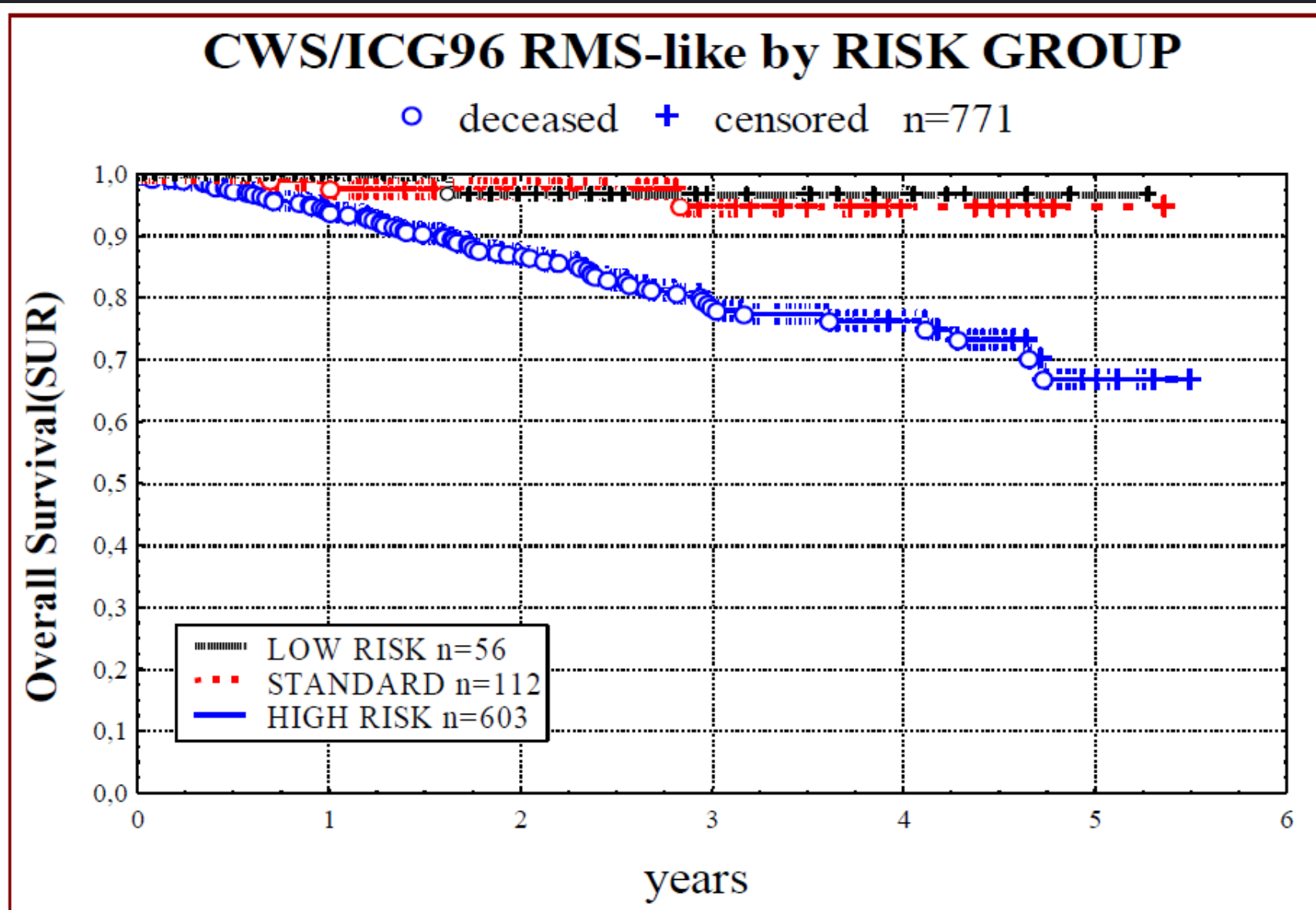


Tobias M. Dantonello et al. JCO 2008;26:406-413

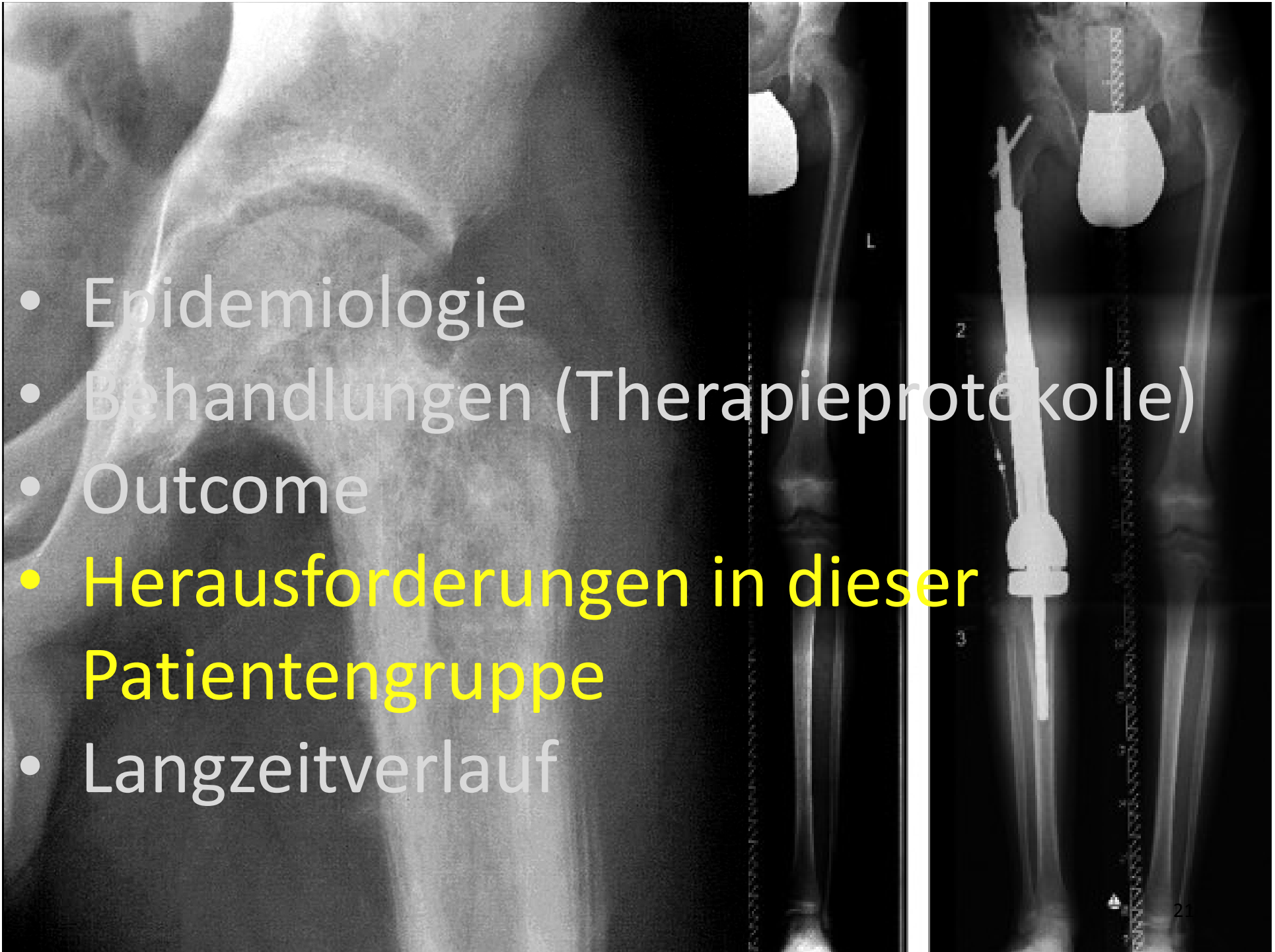
# Resultate der CWS/RMS-96 und MMT-95 Studien



# Resultate der CWS/RMS-96 und MMT-95 Studien



- Epidemiologie
- Behandlungen (Therapieprotokolle)
- Outcome
- Herausforderungen in dieser Patientengruppe
- Langzeitverlauf





# Clinical Oncology in Adolescents and Young Adults

[About Journal](#)[Editors](#)[Peer Reviewers](#)[Articles](#)[View all](#) (24)[Volume 5, 2015](#) (7)[Volume 4, 2014](#) (4)[Volume 3, 2013](#) (8)[Volume 2, 2012](#) (2)[Volume 1, 2011](#) (3)

## Archive: Volume 5, 2015

REVIEW

### Mental health status of adolescent cancer survivors

Mertens AC, Gilleland Marchak J

[Clinical Oncology in Adolescents and Young Adults 2015](#), 5:87-95

Published Date: **18 September 2015**

REVIEW

### Treating melanoma in adolescents and young adults: challenges and solutions

Sreeraman Kumar R, Messina JL, Sondak VK, Reed DR

[Clinical Oncology in Adolescents and Young Adults 2015](#), 5:75-86

Published Date: **15 September 2015**

REVIEW VIDEO

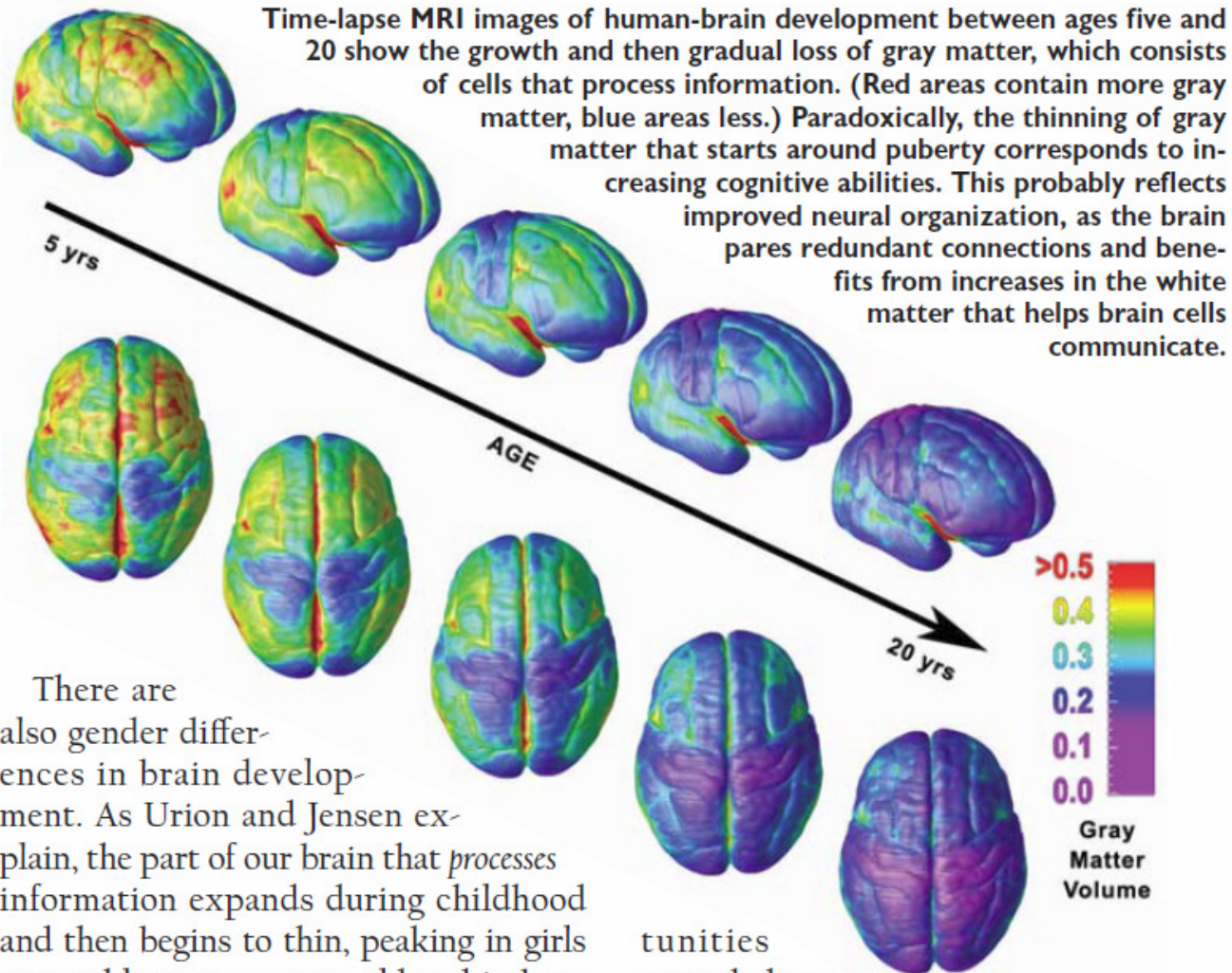
### New strategies to ensure good patient-physician communication when treating adolescents and young adults with cancer: the proposed model of the Milan Youth Project



Magni MC, Veneroni L, Clerici CA, Proserpio T, Sironi G, Casanova M, Chiaravalli S, Massimino M, Ferrari A

[Clinical Oncology in Adolescents and Young Adults 2015](#), 5:63-73

Published Date: **24 August 2015**



There are also gender differences in brain development. As Urien and Jensen explain, the part of our brain that processes information expands during childhood and then begins to thin, peaking in girls at roughly 12 to 14 years old and in boys about two years later. This suggests that girls and boys may be ready to absorb challenging material at different stages, and that schools may be missing oppor-

tunities to reach them.

Meanwhile, the neural networks that help brain cells (neurons) communicate through chemical signals are enlarg-

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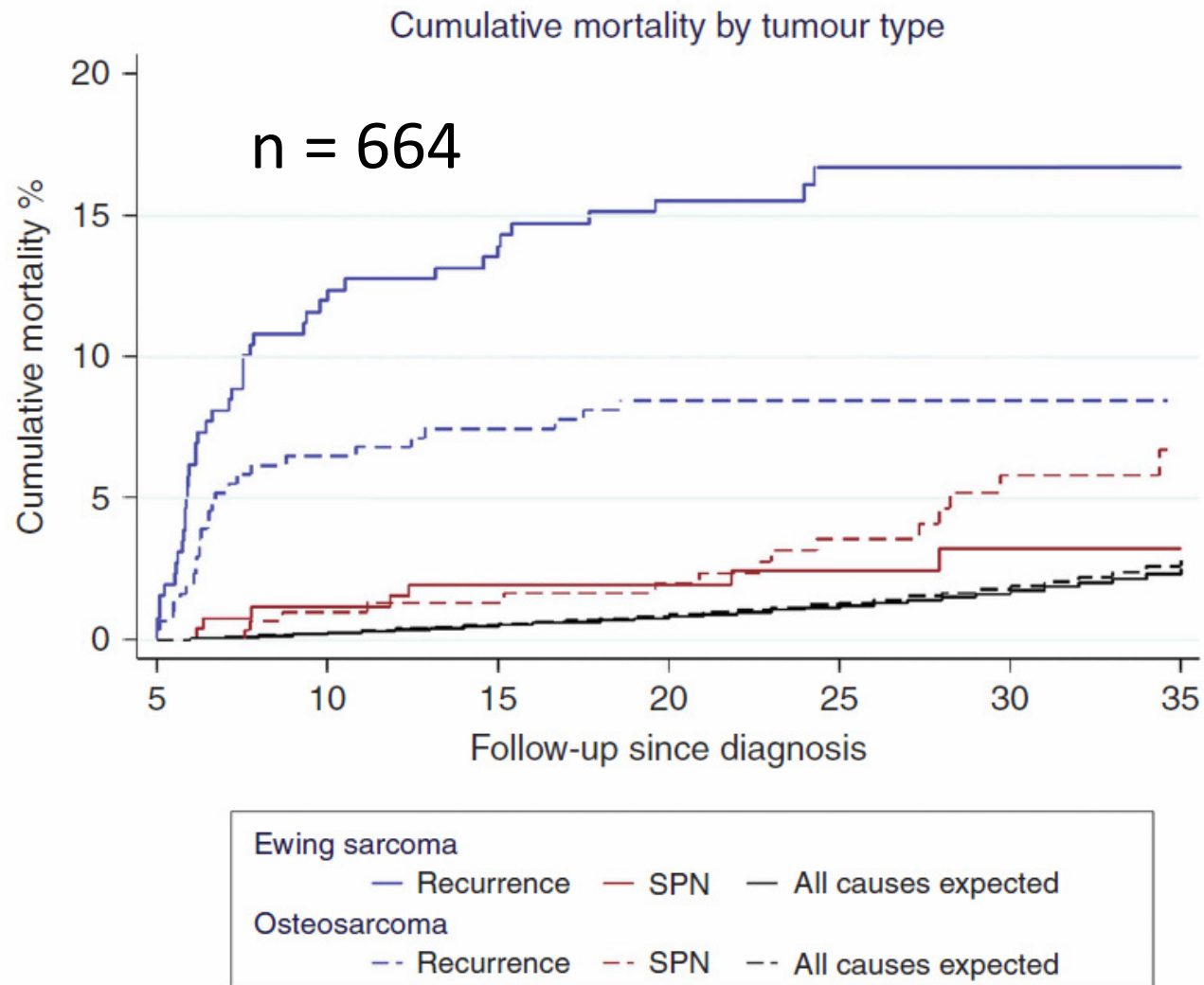
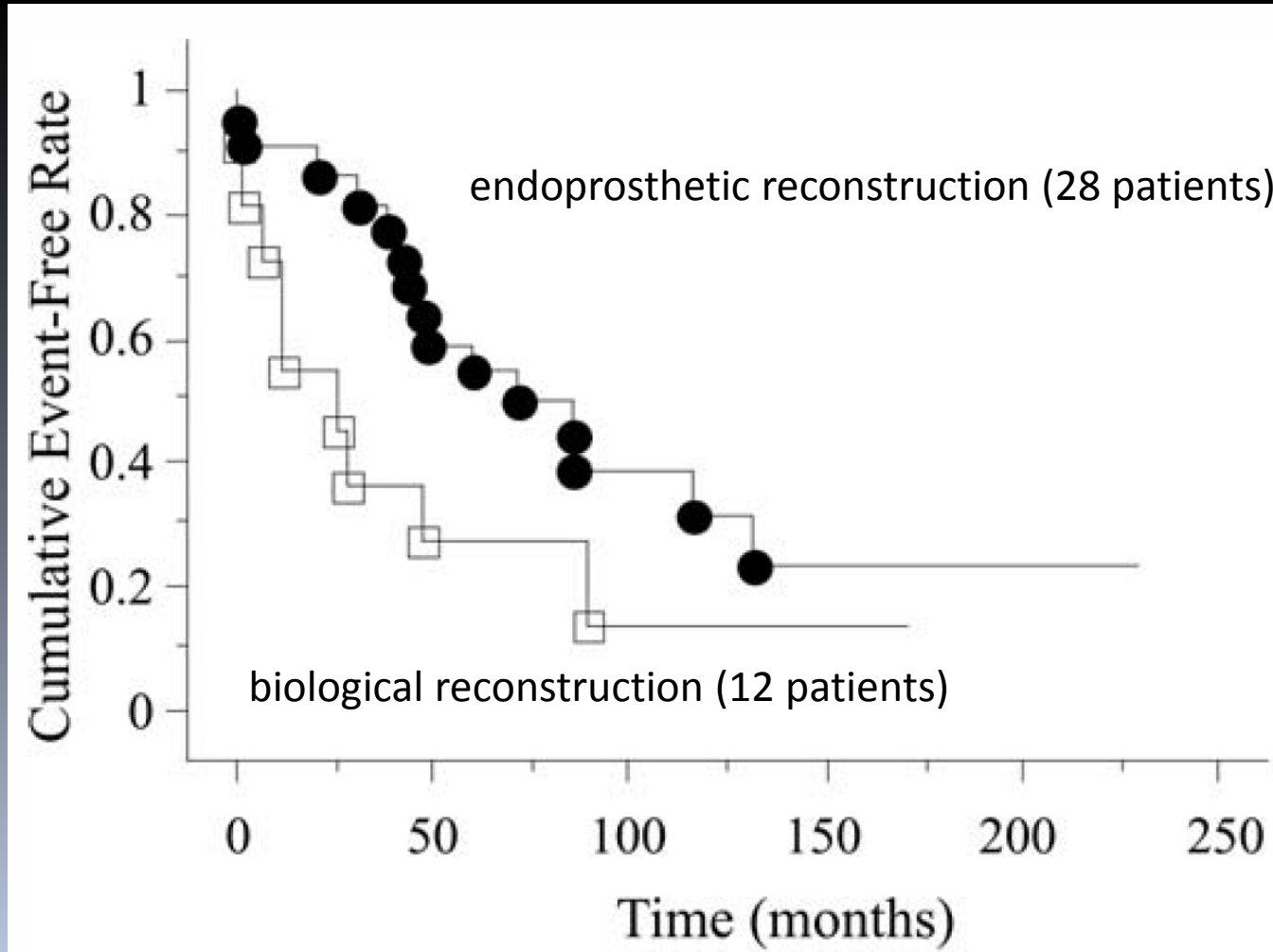


Figure 1. Cumulative mortality of recurrence and second primary neoplasms among childhood bone sarcoma survivors within the British Childhood Cancer Survivor Study (BCCSS) by tumour type.





- (1) 11 jährig oder jünger bei Diagnose
- (2) Primärer maligner Tumor in der Metaphyse und/oder der Epiphyse  
.....des distalen Femurs
- (3) Hatten eine beinerhaltende Operation

# SF-36 health status (n = 664)

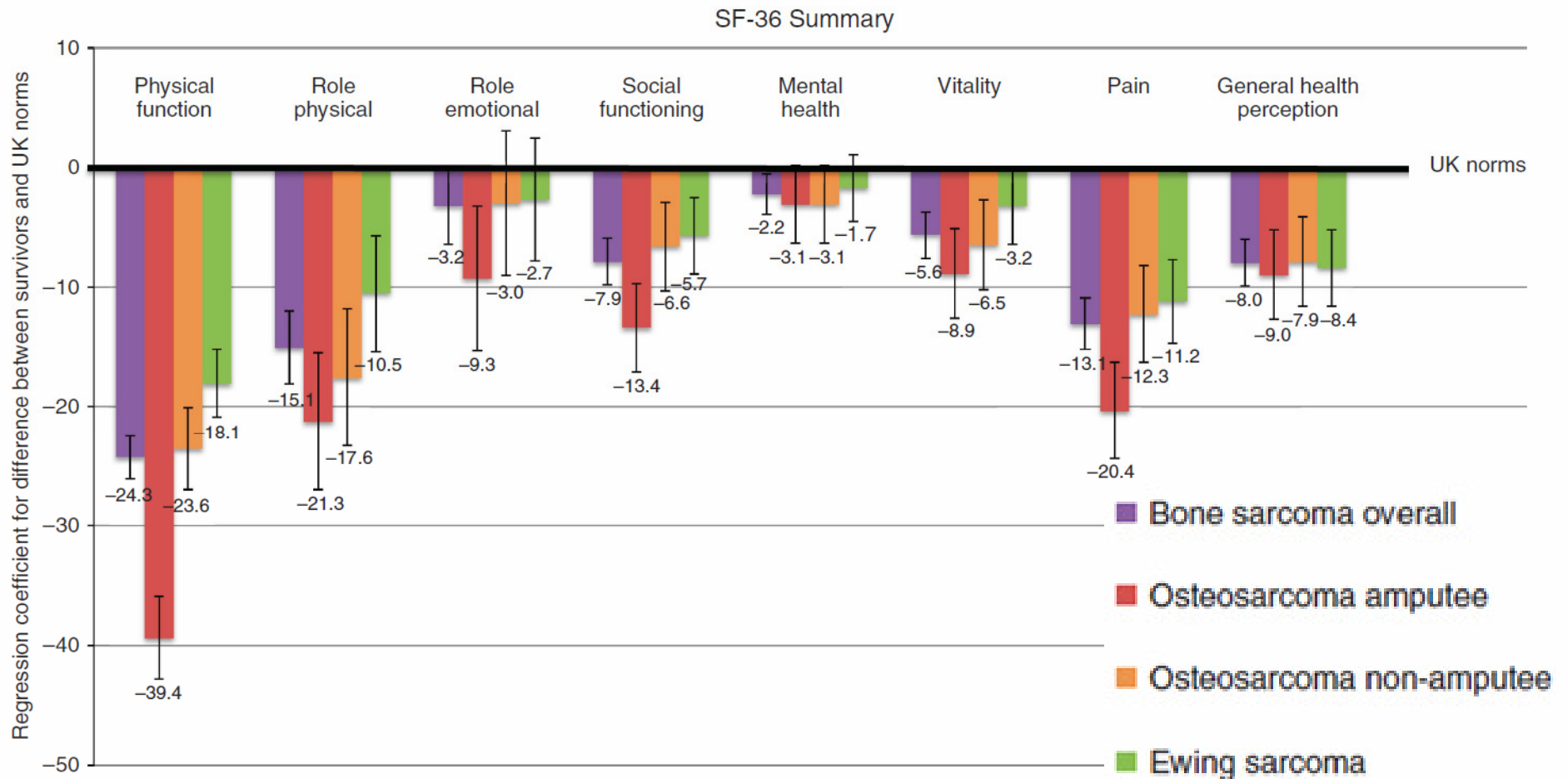
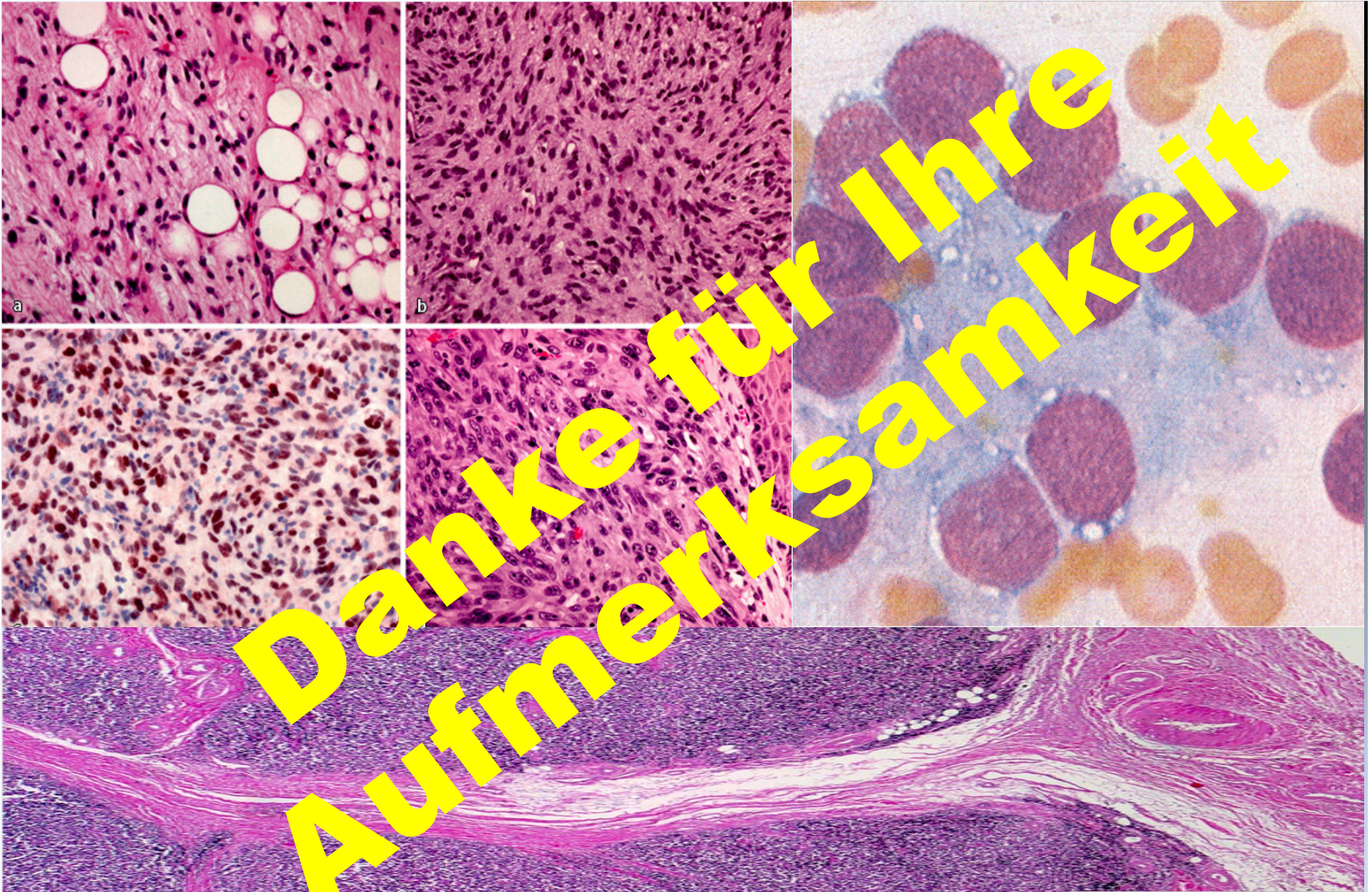


Figure 2. Sex and age adjusted regression coefficients and corresponding 95% confidence intervals for differences in SF-36 health status scales between bone sarcoma, osteosarcoma amputees, osteosarcoma non-amputees, and Ewing sarcoma survivors vs UK norms.





**KINDERKLINIKEN**  
Bern

University Children's Hospital

3. Sarkom-Patiententag, K. Leibundgut, E.M. Tinner

**INSELSPITAL**

UNIVERSITÄTSSPITAL BERN  
HOPITAL UNIVERSITAIRE DE BERNE  
BERN UNIVERSITY HOSPITAL