

MPNST- Surgical Characteristics Of Malignant Peripheral Nerve Sheath Tumour.

Experience Of The Polish Paediatric Solid Tumour Study Group.

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Polish Paediatric Solid Tumour Study Group

Aim of the report

- Aim of the report is to evaluate characteristics and surgical treatment of malignant peripheral nerve sheath tumour (MPNST) in Polish Paediatric Solid Tumour Study Group.

Patients

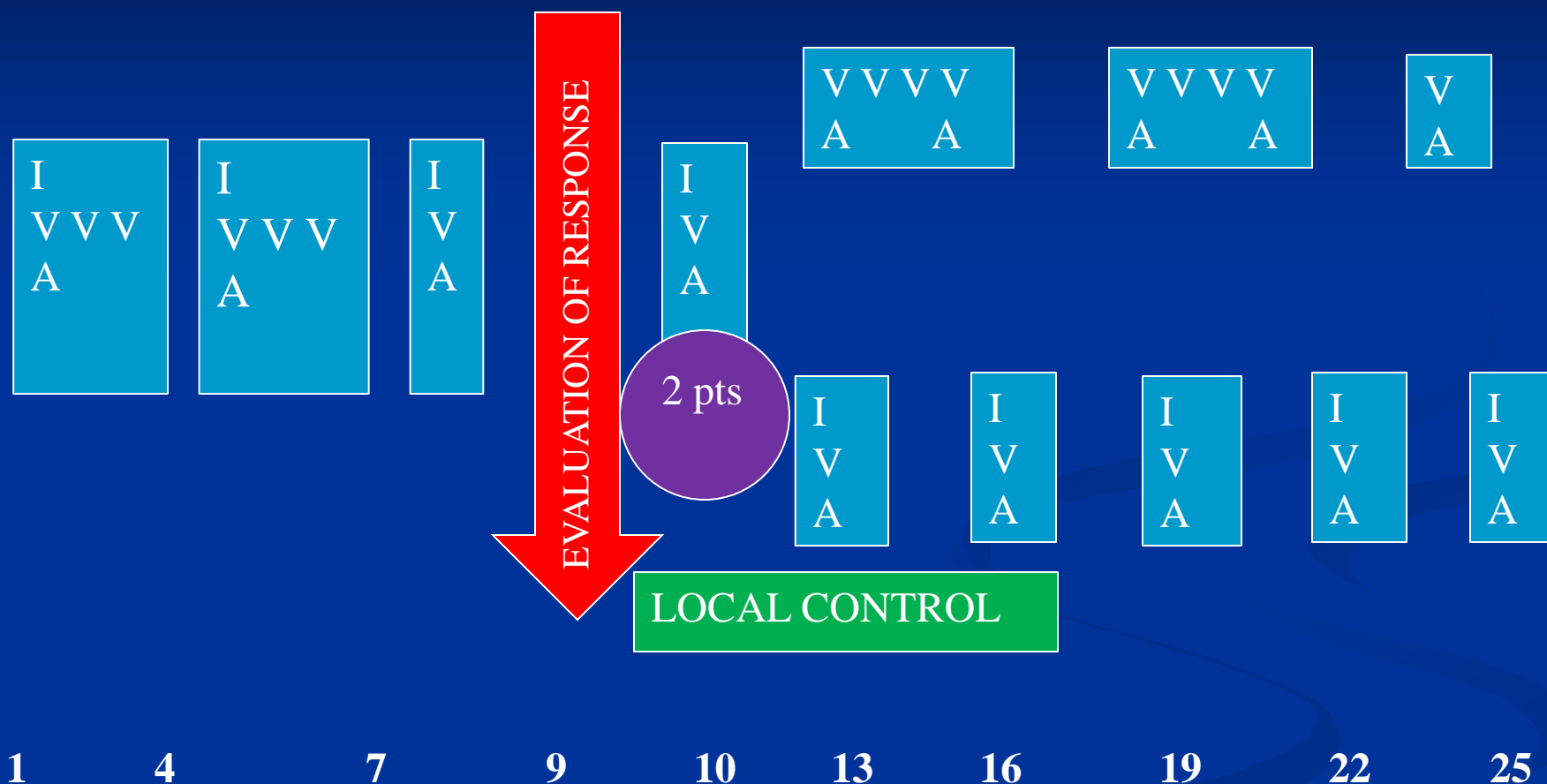
- 381 patients with soft tissue sarcoma staged I-IV were registered and treated in PPSTSG according to CWS 96 and 2002 protocols.
- 12 of them (4/F,8/M) were diagnosed as MPNST and treated according to the non-rhabdomyosarcoma arm in CWS protocols (7/CWS 2002, 5/CWS 96).

Age Mont hs	F/M	Risk grou p	localiza tion	T	N	M	Primar y R	Seco nd look	RTX	CR	relap sed	CR after relap sed	Prog ressi on and died
172	F	HR	Other	2b	0	0	Bx	-	-	-	-	-	+
136	M	HR	Other	2b	0	0	Bx	R2	+	+	+	-	+
140	F	SR	Other	2b	0	0	R2	-	-	-	-	-	+
178	M	SR	Non- PM	1a	0	0	R1	R0	+	SD	+	+	-
180	M	HR	Non- PM	2a	0	1	Bx	R0	+	SD/ CR	-	-	-
209	M	HR	PM	2a	0	0	R2	R1	+	SD	+	-	+
11	F	HR	Other	2b	0	1	R1	-	-	+	-	-	-
171	M	HR	Other	2b	0	1	R2	R0	-	+	-	-	-
178	M	HR	Non- PM	2a	0	0	R2	R0	+	+	-	-	-
36	M	HR	Extre mities	1a	0	0	R1	-	+	+	-	-	-
158	F	HR	Other	2b	0	0	R2	R0	+	+	+	-	+
125	M	HR	Other	2a	0	0	Bx	R1	+	+	-	-	-

Risk group according CWS

- The risk groups (CWS): standard/2, high/10.

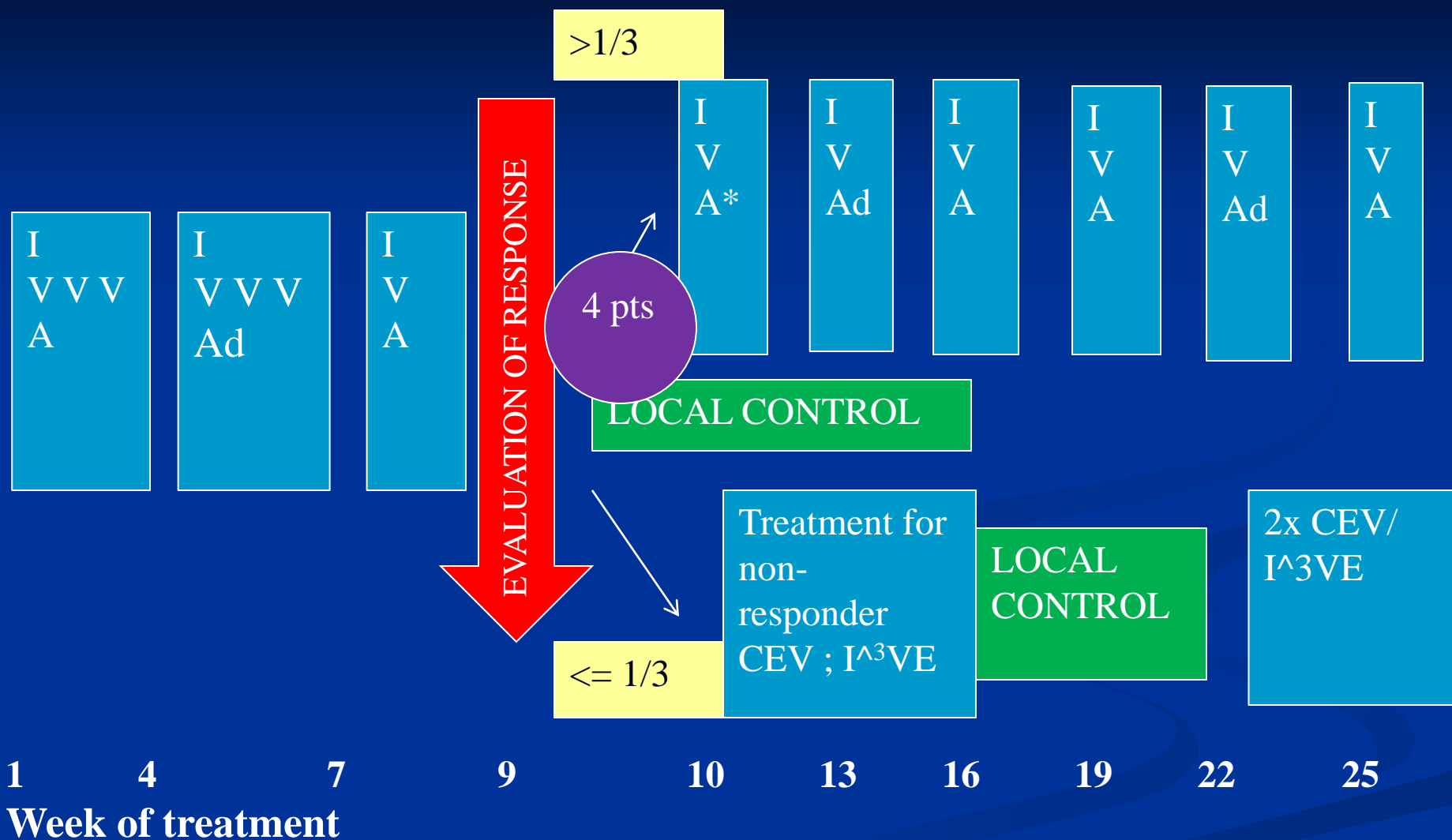
Protocol CWS 2002 – Standard risk group



Week of treatment

I- 2x Ifosfamid 3g/m2; V-1,5mg/m2, Actinomycin 1,5mg/m2

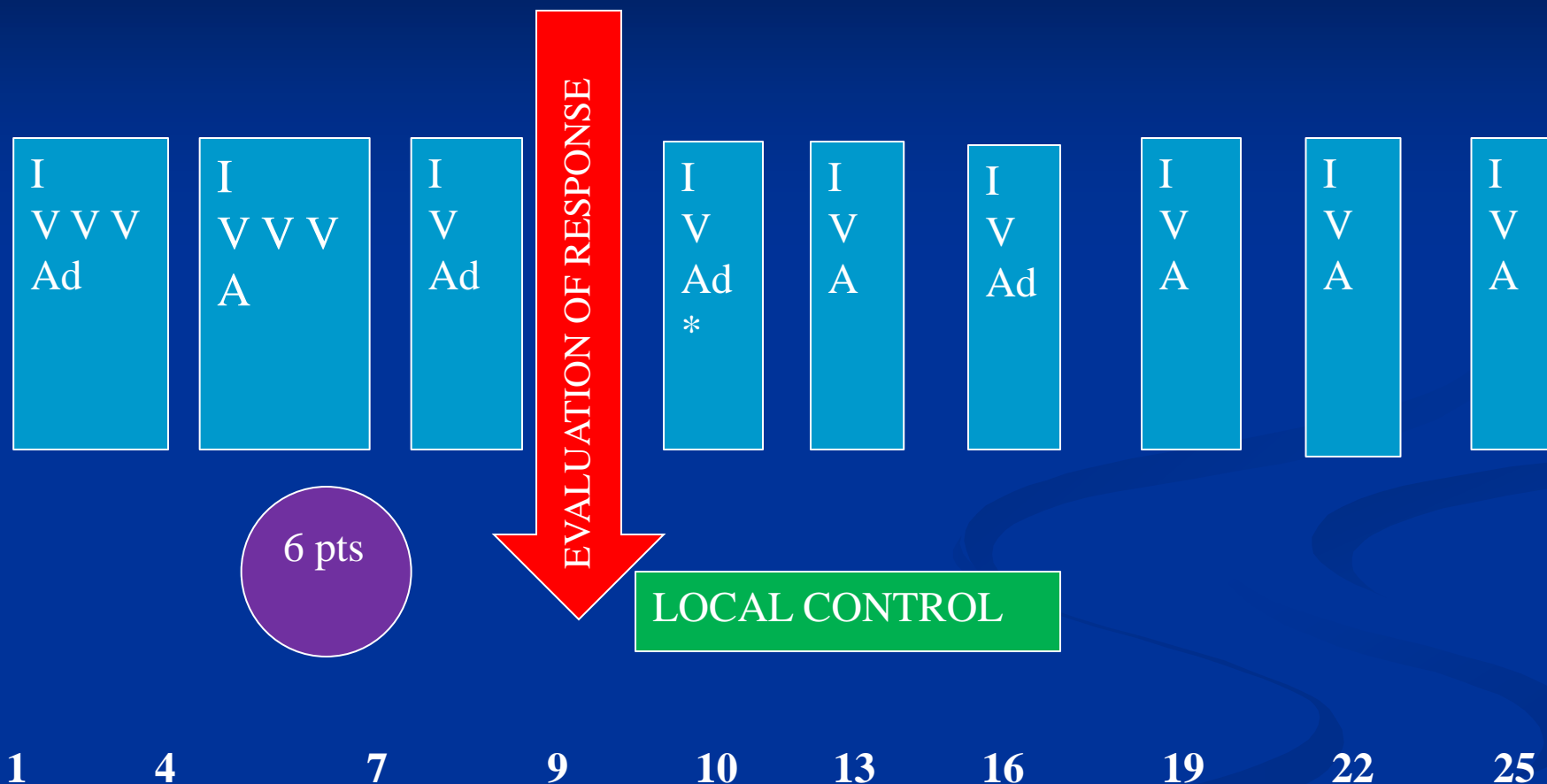
Protocol CWS 96 – High risk group VAIA



I- 2x Ifosfamid 3g/m²; **V-** 1,5mg/m², **A-** Actinomycin 1,5mg/m², **Ad-** 2x Adriamycin 40mg/m², **C-** Carboplatin 500mg/m², **E-** Epirubicin 150mg/m², **E-** Etoposide 150mg/m²

Protocol CWS 2002 – High risk group

VAIA III



6 pts

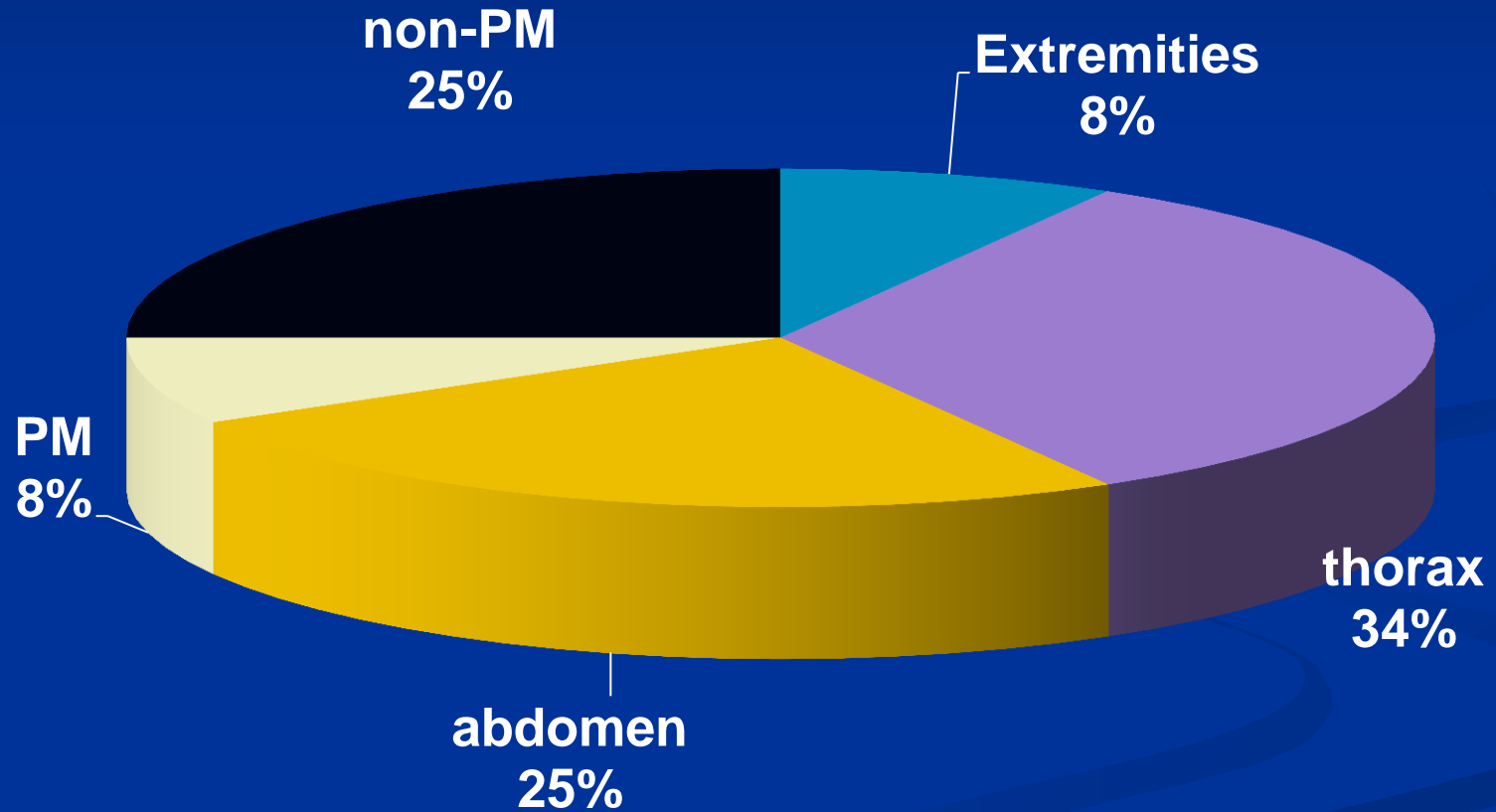
EVALUATION OF RESPONSE

LOCAL CONTROL

Radiotherapy

- 44,8 Gy in both protocol was the same
(CWS 96 and CWS 2002)
SR and HR

Localization of primary tumours



T-status (invasivness)

■ T-status was:

T1a-1, T1b-1,

T2a-4, T2b-5;

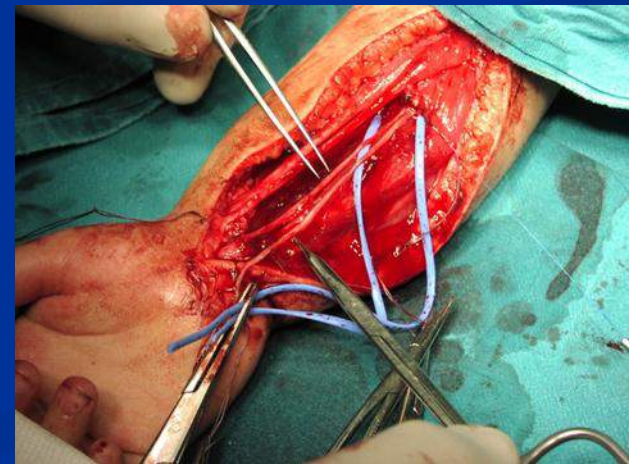
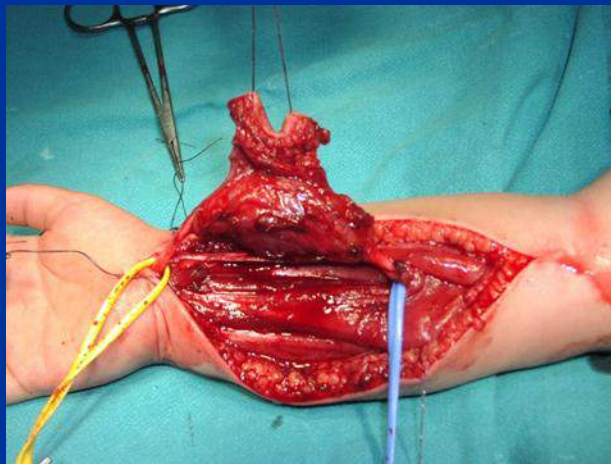
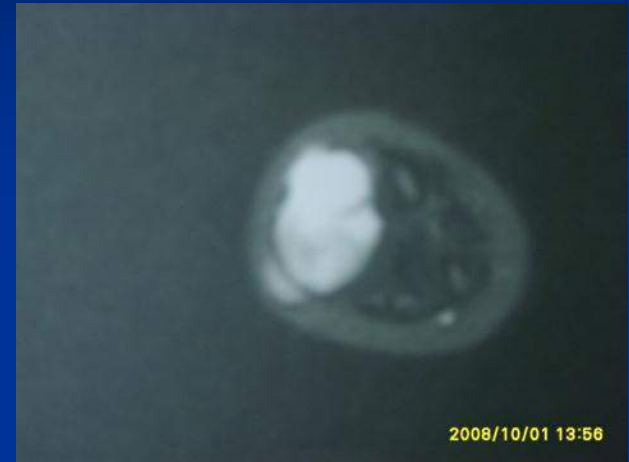
N0-12,

M0-9, M1-3.

Local treatment

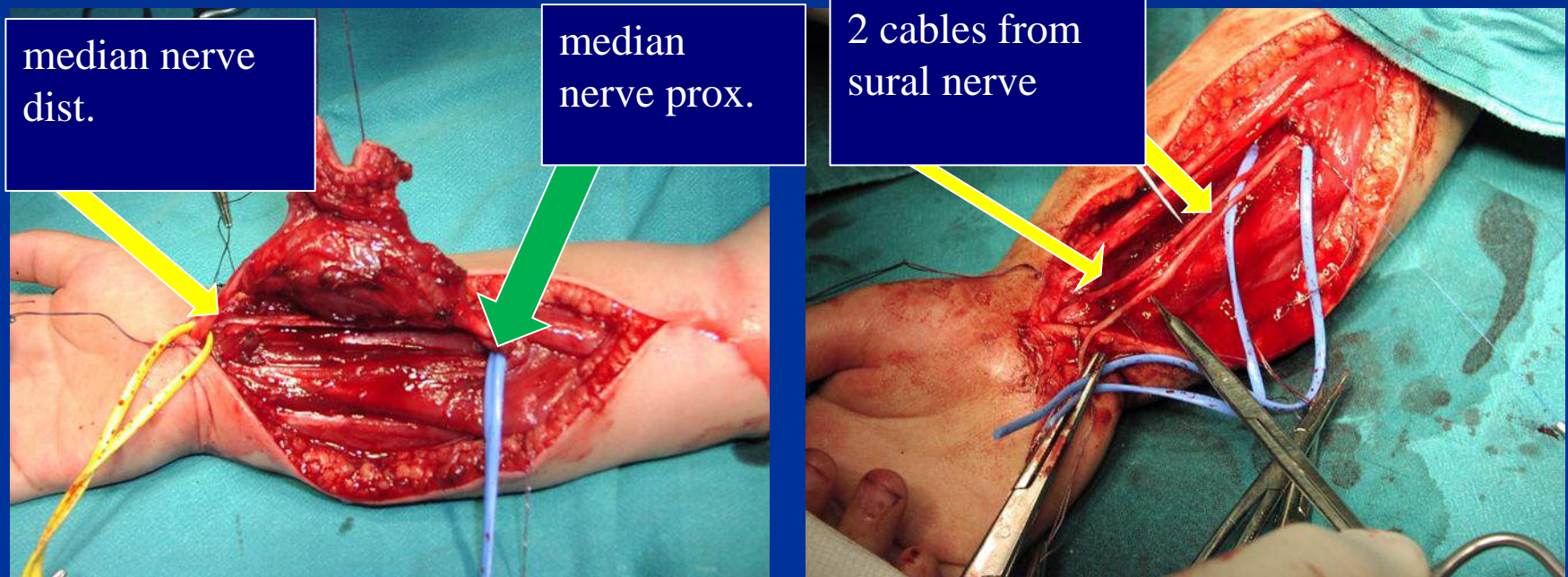
- Consisted of:
 - chemotherapy
 - primary or secondary resection of tumors /R0-0, R1-3, R2-5, biopsy-4/
 - radiotherapy /8 pts/.

MPNST of the median nerve



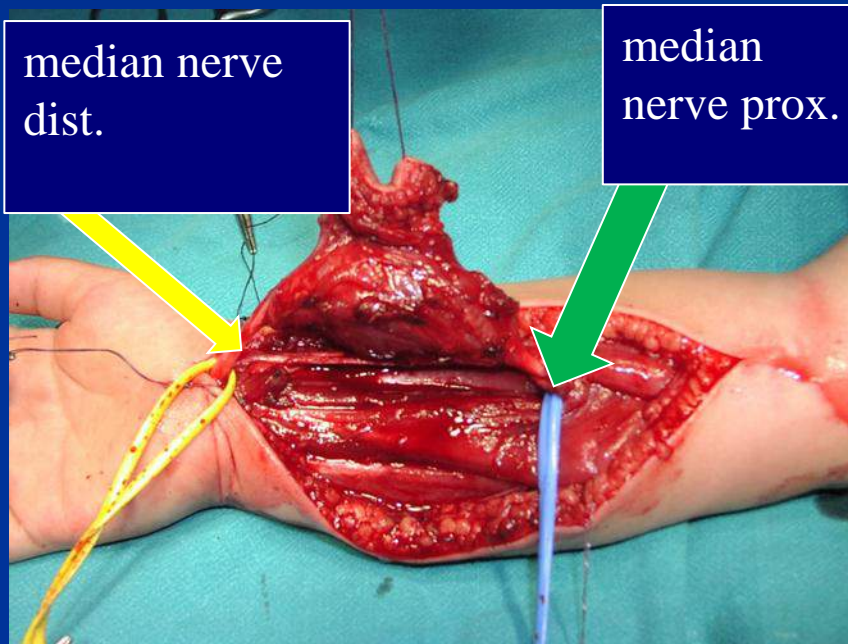
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MPNST

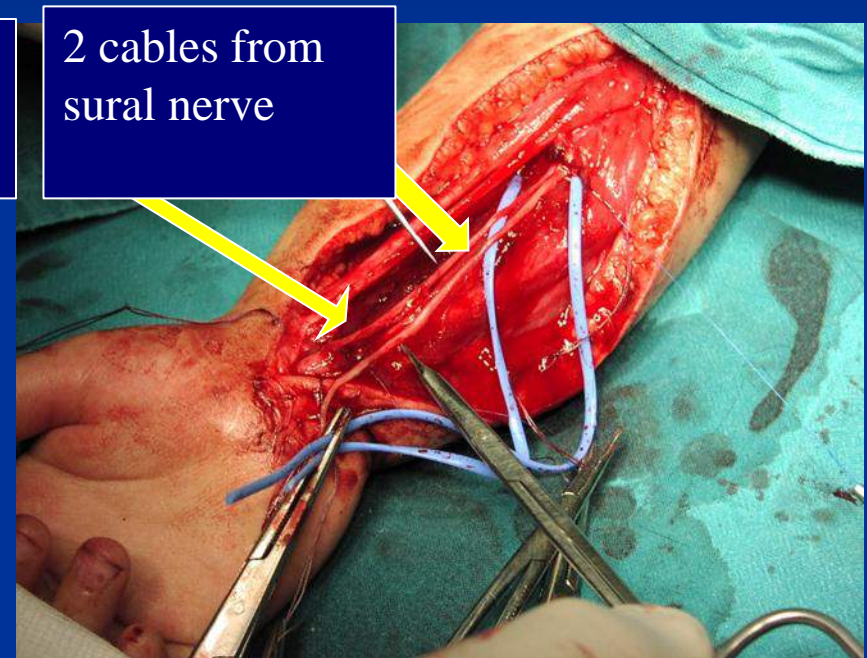


There is no way to avoid loss of function

MPNST



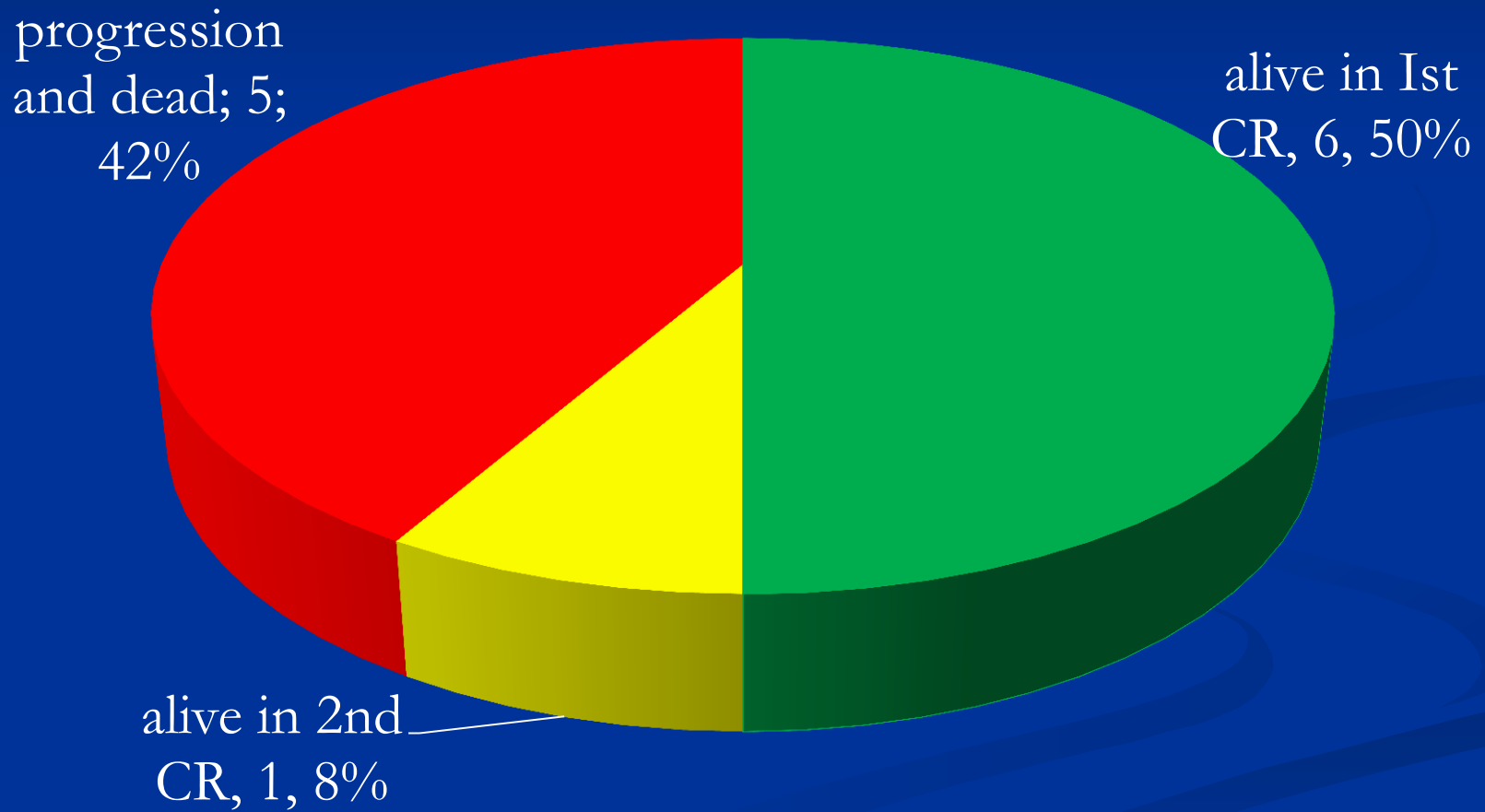
There is no way to avoid loss of function



However, it can be minimized – immediate reconstruction

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Results



Results

- 7 of 12 pts achieved 1st complete remission (CR) after I line treatment
- 4 of 12 pts relapsed (2 after 1st CR, 2 pts with primary SD, achieved CR and later relapsed)
- 5 of 12 pts died
 - 2 pts died in progression of disease (DOD)
 - 3 pt died after relapse (2 pt after primary CR, another with SD, achieved CR , relapsed several times and died)

Results

7/12 pts achieved 1st complete remission (CR) (58,3%) after I line treatment

Number of pt	Primary resection	Secondary resection	Protocol CWS 96/ 2002	RTX	Current status
1	Bx	R2	2002	+	DOD
2	R1	-	2002	-	CR
3	R2	R0	96	-	CR
4	R2	R0	96	+	CR
5	R1	-	96	+	CR
6	R2	R0	96	+	DOD
7	Bx	R1	96	+	CR

Results- regarding primary resection

➤ R1:

2 of 3 pts are alive in 1st CR, 1 pts in 2nd CR after relapse

Results- regarding primary resection

➤ R2:

2 of 5 pts are alive in CR after secondary local Tx

3 of 5 pts died

- *1 pt progressed without local tx*
- *1 pt after SL resection R0 achieved CR, metastatic relapsed after 6 months and died of progression of disease*
- *In 1 case disease was stabilized with chth, pts were submitted to R1 resection, irradiated on and entered CR, subsequently local relapsed and died*

Results- regarding primary resection

➤ Biopsy:

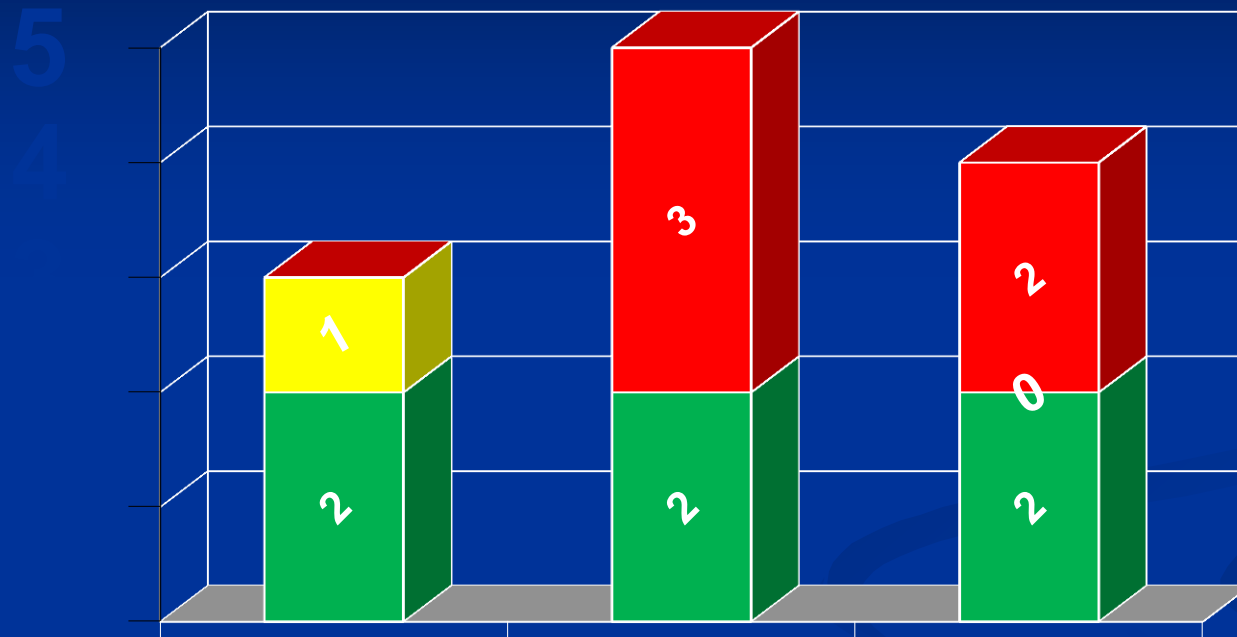
2 of 4 pts are alive in 1st CR after chemotherapy, secondary resection (R0 or R1) and RTX.

2 pts died

1 pt progressed without local tx

1 pt after SL resection R2 achieved CR, metastatic relapsed after 38 months and died of progression of disease

Outcome vs resection of tumour



■ I CR

■ II CR

■ DOD

Conclusion

- Tumors primarily biopsied only and those submitted to unsuccessful surgery (R2) had similar chance for control of the disease after chemotherapy, secondary complete resection (Bx 2/4 vs R2 2/5) and complementary RTX.
- We attribute this finding to biology of MPNST (relatively slow progression and limited invasiveness).

Conclusion

- ? better outcome was observed in patients treated according to CWS 96 protocol then CWS 2002 (4/5 pts in CR 80% vs 4/7 in CR 57,1%).
- ? *difference in total dose of adriamycin in chemotherapy could influenced on the outcome (120 vs 160mg/m²)*
- *Very small numbers (...)*

Thank you for attention

