REVIEW OF LOCAL TREATMENT IN 11 CONSECUTIVE RMS AND RMS-LIKE PATIENTS AGED UNDER 3 YEARS AND TREATED ACCORDING CWS 2002 PROTOCOL.

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Local treatment of youngest patients with soft tissue sarcomas (STS) has well known age-related limits.

Aim of the report is to evaluate the local treatment in group of children aged under 3 years with RMS and RMS-like tumours.
Patients

- Polish Paediatric Solid Tumour Study Group registered 181 patients with STS staged I-IV treated according CWS 2002 protocol.

- 11 pts (1 girl and 10 boys) with RMS and RMS-like tumors (csI-III) younger than 3 years (3-33 mths) were analysed

- Follow-up 67-112 months, M= 90 months.
Types of tumors

- embryonal RMS: 9, 82%
- alveolar RMS: 1, 9%
- PNET-EES: 1, 9%
Locations of primary tumors

- **5, 46%** genito-urinary tract
- **3, 27%** bladder & prostate
- **2, 18%** others
- **1, 9%** extremities

Pie chart showing distribution of primary tumors by location.
T-status; (N0 M0 in all)
Risk grouping according to CWS

- Low: 6, 55%
- Standard: 3, 27%
- High: 2, 18%
Treatment

- chemotherapy /11/
- primary or secondary resection of tumors:
  - R0-5
  - R1-1
  - R2/biopsy- 5
- radiotherapy /4/
Case: 18-month-old girl
eRMS; no response to pre-op ChT amputate?
Case: 18-month-old girl

eRMS; no response to pre-op ChT but in fact the vessels are free
Functional reconstruction: main problem – knee stabilisation/extensin

¼ of the anterior head, and nearly ½ of lateral and medial heads of quadriceps preserved
Results

- 9/11 patients are alive (81.8%):
  - 7 in Ist CR (64%),
  - 2 in PR (18%),
  - 1 pt relapsed (despite primary R0) and died of progression (9%),
  - *1 pt died of sepsis (9%).
4 of 11 pts were irradiated

3 of them are in CR, 1 died in septic shock.
Conclusions

- Favourable outcome in youngest patients seems confirmed.
- Regarding local treatment, supplementary radiotherapy is probably very effective in this group of pts, however toxicity of this treatment must be taken into account.
- Maximal efforts shall be focused on surgical local control of the disease: 4/5 pts who had R0 resection are alive.
Thank you for your attention