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## ABSTRACTS

### **Size does not matter! Predictors of response and survival in desmoplastic small round blue cell tumours (DSRBCT)**

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**Background:** DSRBCT is a rare sarcoma which predominantly affects young men and is associated with a variable response to chemotherapy and poor survival in adults.

**Aims:** We sought to discover whether there were factors which predicted a good response to chemotherapy or improved survival in DSRBCTs.

**Methods:** Review of a prospective database supplemented by retrospective data at a sarcoma referral centre. All histology was reviewed at the sarcoma centre by a sarcoma specialist pathologist.

**Results:** 17 patients were assessed between 1991 and 2006. There were 12 males and 5 females. The median age at presentation was 27 years (range 17-43). 15/17 presented in the retroperitoneum or abdomen, 1 was of a paratesticular origin and the other case was in the upper limb. Median size at presentation was 17.5cm (range 5-21 cm). 15/17 had chemotherapy. Several chemotherapy regimes were used but included ifosfamide and doxorubicin in 14 cases. The other case was given BEP. The median number of cycles of chemotherapy given was 5 (range 1-10). 2 patients had only a single cycle. Radiological evaluation showed 6 partial responses, 5 stable disease and 4 progressive disease. There were no complete responses. Median overall survival (OS) was assessable in 6 cases. The median OS was 12 months but there appeared to be three separate groups of responders: those who progressed and died within 5 months, those who survived the median 12 months and an outlier who survived 62 months. There was no difference in survival or response with age or sex. Neither site nor size of disease at presentation predicted response or survival. Type of chemotherapy and initial response showed the greatest correlation with response and survival.

**Conclusions:** Response to treatment and survival in DSRBCTs is best predicted by initial response to chemotherapy. Differences in survival with different chemotherapy regimens suggest further investigation and clinical trials.