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Unraveling gene fusion dependent transcription factors in Desmoplastic Small Round Cell Tumor

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Desmoplastic small round cell tumor (DSRCT) is a rare and aggressive sarcoma which predominantly occurs in male children and adolescents. Its molecular hallmark is a chromosomal translocation that generates a gene fusion (GF) between *EWSR1*, located on chromosome 22 and *WT1*, located on chromosome 11; a chimeric transcription factor (TF) which is the main driver of the malignant process. Despite aggressive therapy, DSRCT remains a disease with a dismal prognosis with 5-year survival rates around 15%. Current therapeutic approaches are highly toxic and have not achieved complete control of the disease.

Unraveling functional interactions of the GF with other cooperating TF would add further insight into its oncogenic properties to gain deeper knowledge of the biology of this tumor. Hence, in this study we have focused on evaluating the role of TFs that cooperate with the fusion protein in the maintenance of malignancy. In this way, we will be able to define new therapeutic targets and prognostic biomarkers.