Dokumenttyp: journal article

Autor(en) des Beitrags:
Kovar, Heinrich; Amatruda, James; Brunet, Erika; Burdach, Stefan; Cidre-Aranaz, Florencia; de Alava, Enrique; Dirksen, Uta; van der Ent, Wietske; Grohar, Patrick; Grünewald, Thomas G P; Helman, Lee; Houghton, Peter; Iljin, Kristiina; Korsching, Eberhard; Ladanyi, Marc; Lawlor, Elizabeth; Lessnick, Stephen; Ludwig, Joseph; Meltzer, Paul; Metzler, Markus; Mora, Jaume; Moriggl, Richard; Nakamura, Takuro; Papamarkou, Theodore; Radic Sarikas, Branka; Rédini, Francoise; Richter, Guenther H S; Rossig, Claudia; Schadler, Keri; Schäfer, Beat W; Scotlandi, Katia; Sheffield, Nathan C; Shelat, Anang; Snaar-Jagalska, Ewa; Sorensen, Poul; Stegmaier, Kimberly; Stewart, Elizabeth; Sweet-Cordero, Alejandro; Szuhai, Karoly; Tirado, Oscar M; Tirode, Franck; Toretsky, Jeffrey; Tsafou, Kalliopi; Üren, Aykut; Zinovyev, Andrei; Delattre, Olivier

Titel des Beitrags:
The second European interdisciplinary Ewing sarcoma research summit--A joint effort to deconstructing the multiple layers of a complex disease.

Abstract:
Despite multimodal treatment, long term outcome for patients with Ewing sarcoma is still poor. The second “European interdisciplinary Ewing sarcoma research summit” assembled a large group of scientific experts in the field to discuss their latest unpublished findings on the way to the identification of novel therapeutic targets and strategies. Ewing sarcoma is characterized by a quiet genome with presence of an EWSR1-ETS gene rearrangement as the only and defining genetic aberration. RNA-sequencing of recently described Ewing-like sarcomas with variant translocations identified them as
biologically distinct diseases. Various presentations addressed mechanisms of EWS-ETS fusion protein activities with a focus on EWS-FLI1. Data were presented shedding light on the molecular underpinnings of genetic permissiveness to this disease uncovering interaction of EWS-FLI1 with recently discovered susceptibility loci. Epigenetic context as a consequence of the interaction between the oncoprotein, cell type, developmental stage, and tissue microenvironment emerged as dominant theme in the discussion of the molecular pathogenesis and inter- and intra-tumor heterogeneity of Ewing sarcoma, and the difficulty to generate animal models faithfully recapitulating the human disease. The problem of preclinical development of biologically targeted therapeutics was discussed and promising perspectives were offered from the study of novel in vitro models. Finally, it was concluded that in order to facilitate rapid pre-clinical and clinical development of novel therapies in Ewing sarcoma, the community needs a platform to maintain knowledge of unpublished results, systems and models used in drug testing and to continue the open dialogue initiated at the first two Ewing sarcoma summits.