

OUTCOME OF PATIENTS WITH UNDIFFERENTIATED EMBRYONAL SARCOMA OF THE LIVER TREATED ACCORDING TO EUROPEAN SOFT TISSUE SARCOMA PROTOCOLS.

Florent Guérin¹, Hélène Martelli¹, Timothy Rogers², Ilaria Zanetti³, Sheila Terwisscha van Scheltinga⁴, Federica De Corti⁵, Gabriela Guillén Burrieza⁶, Veronique Minard-Colin⁷, Daniel Orbach⁸, Max van Noesel⁴, Marie Karanian⁹, Raquel Davila Fajardo¹⁰, Johannes Merks⁴, Andrea Ferrari¹¹, and Gianni Bisogno³

¹Assistance Publique - Hopitaux de Paris

²University Hospitals Bristol and Weston NHS Foundation Trust

³AIL Padova

⁴Prinses Maxima Centrum voor Kinderoncologie

⁵Universita degli studi di Padova Dipartimento di Geoscienze

⁶Hospital Universitari Vall d'Hebron Servei de Farmacologia Clinica

⁷Institut Gustave Roussy Departement de Medecine Oncologique

⁸Universite Paris Sciences et Lettres Bibliothèques

⁹Centre Leon Berard Departement de Radiotherapie

¹⁰The Ohio State University Department of Radiation Oncology

¹¹Fondazione IRCCS Istituto Nazionale dei Tumori

October 6, 2022

Abstract

Background To assess the outcomes of pediatric patients with Undifferentiated Embryonal Sarcoma of the Liver (UESL) and treatment including at least surgery and systemic chemotherapy. **Methods** This study included patients aged up to 21 years with a pathological diagnosis of UESL prospectively enrolled from 1995 to 2016 in three European trials focusing on the effects of surgical margins, preoperative chemotherapy, use of radiotherapy (RT) and chemotherapy. **Results** Out of 65 patients with a median age at diagnosis of 8.7 years (0.6-20.8), 15 had T2 tumors, and 1 had lymph node spread, 14 were Intergroup Rhabdomyosarcoma Study (IRS) I, 9 IRSII, 38 IRSI, and 4 IRSIV. Twenty-eight upfront surgeries resulted in 5 operative spillages and 11 infiltrated surgical margins, whereas 37 delayed surgeries resulted in no spillages ($P=0.0119$) and 3 infiltrated margins ($P=0.0238$). All patients received chemotherapy, including anthracyclines in 47. Radiotherapy was administered in 15 patients. With a median follow-up of 78.6 months, 5 year overall and event free survivals (EFS) were 90.1% (95%CI 79.2-95.5) and 89.1% (95%CI 78.4-94.6), respectively. Two out of 4 local relapses had previous infiltrated margins and 2 out of 3 patients with metastatic relapses received reduced doses of alkylating agents. Infiltrated margins ($P=0.1607$), T2 stage ($P=0.3870$), use of RT ($P=0.8731$), and anthracycline-based chemotherapy ($P=0.1181$) were not correlated with EFS. **Conclusions** Neoadjuvant chemotherapy for pediatric patients with UESL increases the probability of complete surgical resection. The role of anthracyclines and radiotherapy for localized disease remains unclear. The use of alkylating agents is recommended.

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