

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

Florent Guérin<sup>1</sup>, Hélène Martelli<sup>1</sup>, Timothy Rogers<sup>2</sup>, Ilaria Zanetti<sup>3</sup>, Sheila Terwisscha van Scheltinga<sup>4</sup>, Federica De Corti<sup>5</sup>, Gabriela Guillén Burrieza<sup>6</sup>, Veronique Minard-Colin<sup>7</sup>, Daniel Orbach<sup>8</sup>, Max van Noesel<sup>4</sup>, Marie Karanian<sup>9</sup>, Raquel Davila Fajardo<sup>10</sup>, Johannes Merks<sup>4</sup>, Andrea Ferrari<sup>11</sup>, and Gianni Bisogno<sup>3</sup>

<sup>1</sup>Assistance Publique - Hopitaux de Paris

<sup>2</sup>University Hospitals Bristol and Weston NHS Foundation Trust

<sup>3</sup>AIL Padova

<sup>4</sup>Prinses Maxima Centrum voor Kinderoncologie

<sup>5</sup>Universita degli studi di Padova Dipartimento di Geoscienze

<sup>6</sup>Hospital Universitari Vall d'Hebron Servei de Farmacologia Clínica

<sup>7</sup>Institut Gustave Roussy Département de Médecine Oncologique

<sup>8</sup>Université Paris Sciences et Lettres Bibliothèques

<sup>9</sup>Centre Leon Berard Département de Radiothérapie

<sup>10</sup>The Ohio State University Department of Radiation Oncology

<sup>11</sup>Fondazione 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 IRSIII, 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.

## Hosted file

ueslv2.0.docx available at <https://authorea.com/users/404146/articles/589239-outcome-of-patients-with-undifferentiated-embryonal-sarcoma-of-the-liver-treated-according-to-european-soft-tissue-sarcoma-protocols>

