



Combined in situ hypothermic liver preservation and cardioplegia for resection of hepatoblastoma with intra-atrial extension in a 3 year old child



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ABSTRACT

Cure of hepatoblastoma requires complete macro- and microscopic resection of the tumor, without tumor rupture. In case of hepatoblastoma with intra-atrial tumor extension (ITE), “en bloc” resection of the hepatic tumor and ITE, with minimal risk of postoperative liver failure, constitutes a surgical challenge. We report on a 3 year old child with hepatoblastoma of the right liver lobe, and ITE through the upper Inferior Vena Cava. Initial chemotherapy (SIOPEL IV HR) induced good response, but tumor persisted inside the right atrium with tight adhesions to the cardiac wall. “En bloc” right extended hepatectomy and removal of the ITE with reconstruction of the atrial and caval wall with autologous pericardial patch was performed under normothermic extracorporeal circulation and cardioplegia, combined with in situ hypothermic liver preservation of the remaining left liver. Complete tumor resection was achieved without tumor rupture. Postoperative liver function was immediately good and adjuvant chemotherapy was resumed per protocol. Eleven months after the end of treatment the child is in complete tumor remission. In children with hepatic tumor and ITE, combination of normothermic extracorporeal circulation with cardioplegia and in situ hypothermic liver preservation allows “en bloc” extended hepatectomy and removal of ITE, with limited risk of postoperative liver failure.

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The aim of surgery for hepatoblastoma (HB) is complete resection of the tumor without rupture. If the tumor has an intra-atrial tumor extension (ITE), combination of hepatobiliary and cardiac surgical techniques is needed. In case of extended hepatectomy leaving a small liver remnant, normothermic vascular exclusion exposes the patient to the risk of postoperative acute liver failure, which may lead to urgent liver transplantation (LT). To reduce this

risk and allow complex liver resections, three techniques of hypothermic liver preservation (HLP) have been described: in situ, ante situm and ex situ HLP.

The aim of this article is to report on our experience of “en bloc” resection of a large HB of the right liver with ITE in a 3-year-old child combining in situ HLP and cardioplegia under normothermic extracorporeal circulation (ECC).

1. Case report

1.1. Case presentation

A 32-month-old male was presented with abdominal pain and vomiting. Physical examination revealed a large abdominal mass. Imaging with Ultrasound-Doppler (US) and computed tomography

Abbreviations: AA, Ascending Aorta; CT, computed tomography; ECC, extracorporeal circulation; HB, hepatoblastoma; HLP, hypothermic liver preservation; ITE, intra-atrial tumor extension; IVC, Inferior Vena Cava; LT, liver transplantation; SIOPEL, International Childhood Liver Tumors Strategy Group of the SIOP; Société Internationale d'Oncologie Pédiatrique; SVC, Superior Vena Cava; US, ultrasound.

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(CT) showed a large hepatic tumor of the right liver located in segments V, VI, VII and I measuring $9.4 \times 9.0 \times 12.8$ cm, and a small satellite lesion of 1.2 cm diameter in segment V (Fig. 1). The main tumor extended into the right hepatic vein, the Inferior Vena Cava (IVC) and the right atrium, where its size was 3.5×3 cm. Its left edge touched the median hepatic vein. Portal branches were free of tumor. There was no intraperitoneal rupture and no distant metastasis. Alphafetoprotein level was $384\,000 \mu\text{g/L}$. Percutaneous ultrasound guided needle core biopsy confirmed the diagnosis of HB. Neither portal, nor central venous emboli were observed. The radiological staging was PRETEXT II V3 according to the International Childhood Liver Tumors Strategy Group (SIOPEL) staging [1]. The patient was treated according to the SIOPEL IV high risk protocol, using cisplatin and doxorubicin [2]. Due to good tumor response and poor tolerance to chemotherapy, the third preoperative cycle was skipped.

1.2. Situation at surgery and description of intervention

Eleven weeks after diagnosis CT showed a significant reduction of the main tumor with an estimated volumetric reduction of 90% (Fig. 1). Nevertheless, tumor persisted in the right atrium with tight contact and possible invasion of its posterior wall. Alphafetoprotein markedly decreased ($168 \mu\text{g/L}$ at day of surgery). The remaining liver free of tumor included segments II, III, left $2/3$ of IV and left half of I, with left vessels. Its mass was estimated by CT volumetry between 200 and 250 g, representing 1.5–1.8% of the child’s body mass.

The operation took place 13 weeks after diagnosis at the age of 35 months (weight 13.35 kg) and can be watched in Video 1. It was carried out through a median sternotomy, prolonged by an inverted T laparotomy. In situ HLP was prepared as follows (Fig. 2): 1) the inferior mesenteric vein was cannulated up to the splenomesenteric confluence; 2) the lower IVC was cannulated up to the renal veins; 3) the right branch of the portal vein was cut and a cannula was inserted in its proximal segment toward the left portal vein; 4) the infrahepatic IVC and the suprahepatic IVC were isolated. The hepatectomy started without vascular exclusion of the liver at the right border of the common trunk of the median and left hepatic veins, preserving the left $2/3$ of segment IV, and progressed toward

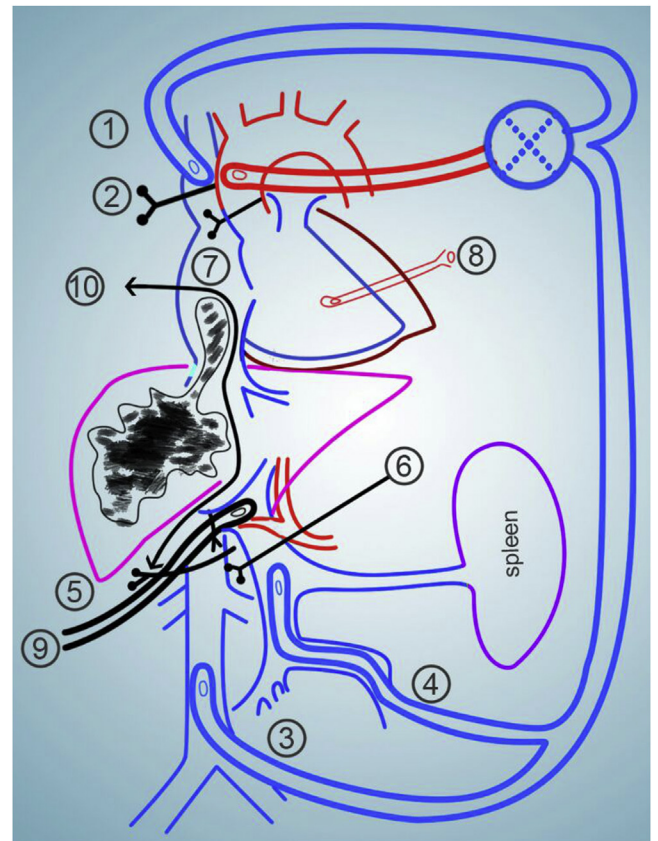


Fig. 2. Operation scheme. 1. SVC cannula, 2. SVC clamp, 3. IVC cannula, 4. Portomesenteric cannula, 5. Infra-hepatic suprarenal IVC clamp, 6. Hepatic pedicle clamp (Pringle's maneuver), 7. Right atrium opening for heart and liver remnant drainage, 8. Left atrium drainage cannula, 9. Portal cannula (through right portal stump) for in situ liver remnant perfusion of 4°C preservation solution, 10. "En bloc" hepatectomy and ITE resection line.

the origin of the right portal pedicle. The lower retrohepatic IVC was freed from the right liver, which remained attached only by the right hepatic vein and the ITE (Fig. 3). The superior vena cava (SVC)

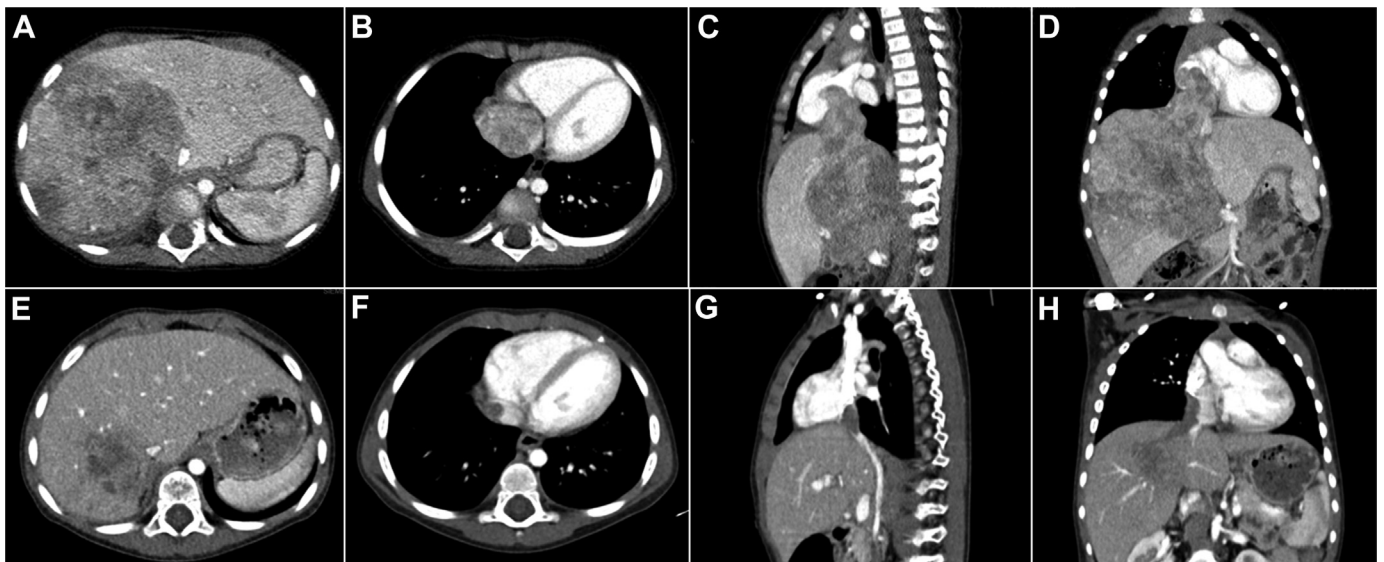


Fig. 1. Radiological presentation on injected CT of the tumor at diagnosis (A–D) and after 2 courses of chemotherapy (SIOPEL IV HR) at the time of surgery (E–H). Size at diagnosis was $94 \times 90 \times 128$ mm for the main liver tumor and 36×28 mm for the tumor in the right atrium; pre-operatively the main tumor was reduced to $48 \times 40 \times 49$ mm and the tumoral extension to the IVC and the right atrium to $11 \times 11 \times 27$ mm.

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