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## Review

# Chordoma in children: Case-report and review of literature



Jean-Louis Habrand<sup>a,b,c,\*</sup>, Jean Datchary<sup>a,b,c</sup>, Stéphanie Bolle<sup>c</sup>,  
Anne Beaudré<sup>c</sup>, Ludovic de Marzi<sup>b</sup>, Kévin Beccaria<sup>d</sup>, Dinu Stefan<sup>a</sup>,  
Jacques Grill<sup>e</sup>, Rémi Dendale<sup>b</sup>

<sup>a</sup> Department of Radiation Oncology, Centre François Baclesse, 3 rue du Général Harris, 14076 Caen, France

<sup>b</sup> Department of Radiation Oncology, Institut Curie Protontherapy Center, Campus universitaire, 91406 Orsay, France

<sup>c</sup> Department of Radiation Oncology, Gustave Roussy Cancer Campus, 114 rue Edouard Vaillant, 94805 Villejuif, France

<sup>d</sup> Department of Pediatric Neurosurgery, Centre Hospitalier Necker-Enfants Malades, 149 rue de Sèvres, 75015 Paris, France

<sup>e</sup> Department of Pediatric Oncology, Gustave Roussy Cancer Campus, 114 rue Edouard Vaillant, 94805 Villejuif, France

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## ABSTRACT

We report an exceptional case of a very late local failure in a 9-year-old boy presenting with a chordoma of the cranio-cervical junction. The child was initially treated with a combination of surgical resection followed by high dose photon–proton radiation therapy. This aggressive therapy allowed a 9-year remission with minimal side-effects. Unfortunately, he subsequently presented with a local failure managed with a second full-dose course of protons. The child died one year later from local bleeding of unclear etiology.

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## 1. Introduction

Chordomas (CH) are rare low grade malignancies that represent approximately 1% of all intra cranial tumors and 4% of bony primaries.<sup>1–3</sup> They develop from notochordal embryonic residues,<sup>4</sup> an origin supported by the association with the brachyury gene, a gene involved in the notochord

development.<sup>5,6</sup> In the adults, they are located typically in the sacrococcygeal (SC) (50%), the intra cranial (IC), at the skull base (SB) (35%), and the intermediate spinal (S) regions (15%). Only 5% of cases have been described in the pediatric age, with unclear specificities concerning presentation, and outcome. We report in this paper a clinical case of skull base CH in a child, and we summarize through the English literature peculiarities of CH in this age group.

\* Corresponding author at: Department of Radiation Oncology, Centre François Baclesse, 3 rue du Général Harris, 14076 Caen, France. Tel.: +33 231455020; fax: +33 231455069.

E-mail address: [France.jl.habrand@baclesse.unicancer.fr](mailto:France.jl.habrand@baclesse.unicancer.fr) (J.-L. Habrand).

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## 2. Case report

Antonio M., a 9-year-old boy, was referred to the Department of Neurosurgery in San Giovanni Rotondo (Italy), following a couple of month-history of posterior cervical pains. On MRI, an extensive tumor process was visualized. It extended from the lower clivus, through the foramen magnum, down to C2, inclusive. The tumor was abutting the medulla and cervical cord anteriorly and laterally, right side. It extended substantially in the soft tissues anteriorly, with a visible protrusion, on physical examination, through the posterior aspect of the nasopharynx. Estimated dimensions were 35 mm × 15 mm. The child was operated on through two successive approaches, in March and April 2000. The tumor bed was filled with fatty and muscular tissue. Pathological examination was consistent with a CH. On histopathological study, the tumor was positive for epithelial (KL1+, EMA+), and mesenchymal (PS100) antigens. Post-operative MRI was of a difficult interpretation, due to the interposition of soft tissue although multiple small residual foci were suspected within the canal. The patient was referred to the Institut Gustave-Roussy, in Villejuif (France) where high dose, high precision post-operative radiotherapy was recommended. A combined photon-protontherapy program was implemented in collaboration with the Institut Curie-Proton Therapy Center, in Orsay (ICPO, France). It

delivered, from January 2001 through March 2001, a total of 68.4 Gy (RBE) in 5 daily fractions of 1.8 Gy each per week (Gy (RBE) corresponds to the physical dose times an estimated 1.1 mean RBE value). Half was delivered using 3D conformal 15 MV photons, and half using 201 MeV protons, with a fixed horizontal beam, on a passive scattering mode (Fig. 1). Acute tolerance was satisfactory with mild headaches, nausea, and mucositis, managed with a short course of steroids and mouthwash. Follow-up was alternated between France and Italy and uneventful until January 2010. At that time, 106 months following radiotherapy, performance status was excellent, with only a slight permanent neck deviation, related with mild radiation-induced atrophy of C1, and of the occipital bone (Fig. 2). C1 deformation was attributed retrospectively to left to right vertebral body dose-gradient that ranged between 9 and 48 Gy. Unfortunately, MRI also evidenced a local tumor progression along the pharyngeal wall. A grossly subtotal resection was performed abroad in the same neurosurgical department, in May 2010, followed by a second course of conventionally fractionated protons, at ICPO, up to 70 Gy (RBE) (Fig. 3). The cumulative biologically equivalent dose (BED) was estimated retrospectively to 165 Gy<sub>10</sub> for tumor, and 270 Gy<sub>2</sub> for late CNS reactions. These theoretical estimates do not take into account the extensive delay between treatments. The patient could resume normal life for almost a year, when a cataclysmic hemorrhage was exteriorized through the mouth.

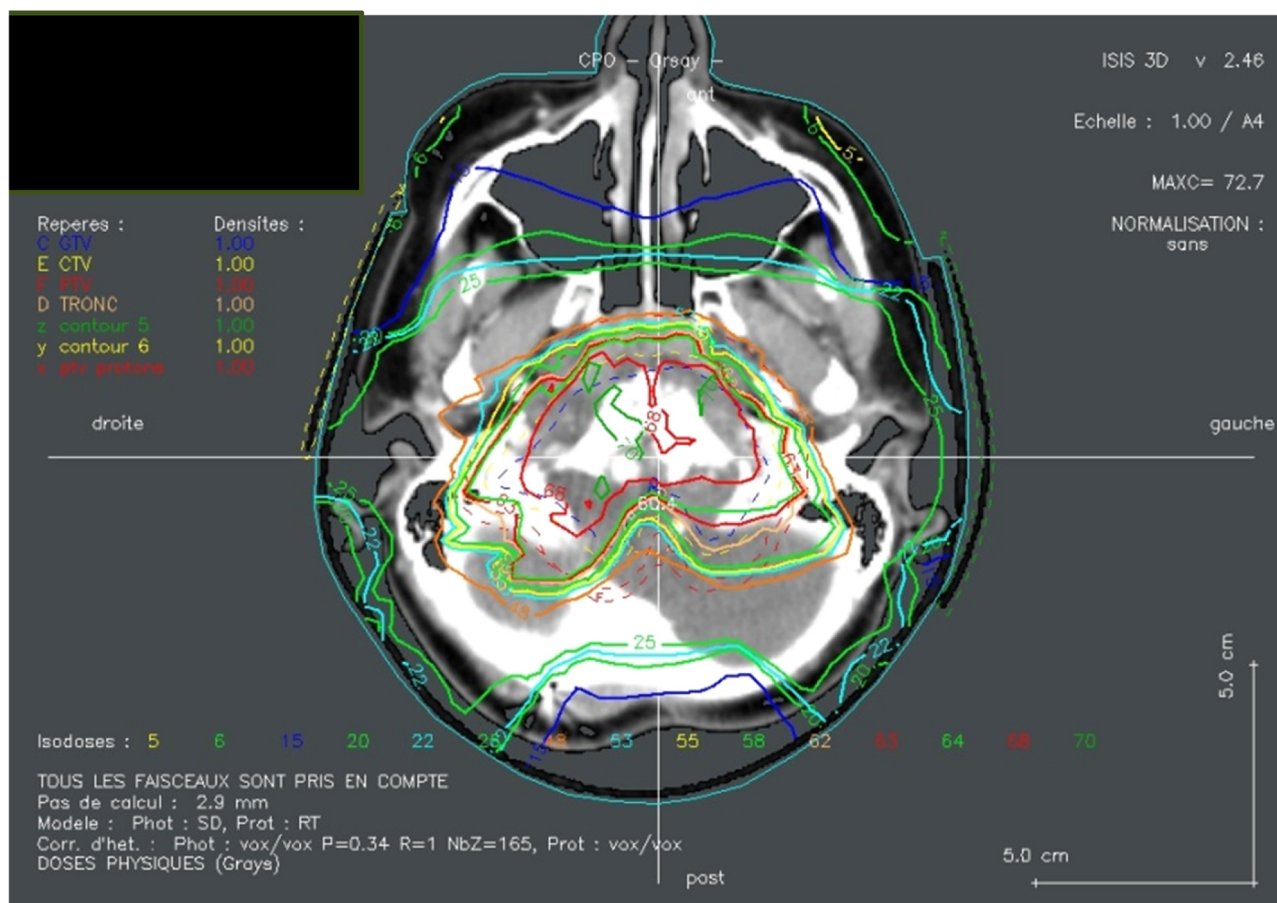


Fig. 1 – 9 year-old boy with skull base-cervical canal chordoma. Post-operative photon-proton radiotherapy mid-plane dose-distribution. Prescribed dose: 68.4 Gy (RBE) (personal coll.).

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