Pancreatoblastoma in children: EXPeRT/PARTNER diagnostic and therapeutic recommendations.

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Abstract

Pancreatoblastoma (PBL) is a rare malignant epithelial neoplasm affecting typically young children. Signs related to advanced upper-abdominal tumor accompanied by elevated serum ∝-fetoprotein levels in a young child suggest PBL, however histopathological examination is required for diagnosis. The mainstay of treatment is a complete surgical resection. Inoperable and/or metastatic PBL may become amenable to complete, delayed surgery after neo-adjuvant chemotherapy. This manuscript presents the internationally consensus recommendations for the diagnosis and treatment of children with PBL, established by the European Cooperative Study Group for Pediatric Rare Tumors (EXPeRT) within the EU-funded PARTNER (Paediatric Rare Tumors Network – European Registry) project.

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`Table 1. Current classification of pancreatic cancers based on the 8th edition of the AJCC Cancer Stagi`

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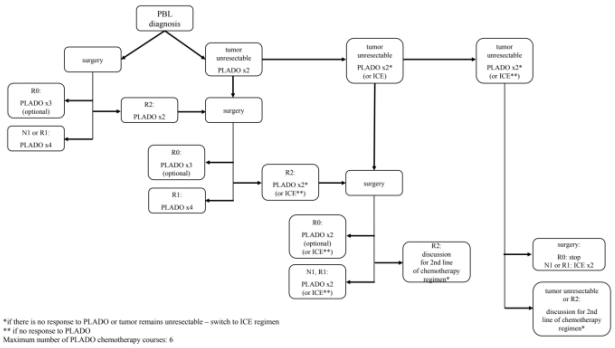
Table 2. Staging system for PBL based on the results of initial surgery proposed by EXPeRT.pdf available at https://authorea.com/users/397637/articles/510460-pancreatoblastoma-in-children-expert-partner-diagnostic-and-therapeutic-recommendations



Abbreviations : PLADO : platinum + doxorubicin chemotherapy, R0 : complete exision, R1 : microspic residues, R2 : macroscopic residues, , ICE : ifosfamide carboplatin and etoposide, N1 – involved Lymph nodes

Figure 1. Proposal strategy from EXPERT for the treatment of pediatric pancreatoblastoma.