Pancreatoblastoma: A report from the European cooperative study group for paediatric rare tumours (EXPert)

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- Joint analysis of all cases registered between 2000 and 2009 by the national groups of Italy, France, United Kingdom, Poland and Germany

- 20 patients <18 years (median 4)
  - 9 had distant metastases at diagnosis
  - 17 had tumour resection, at initial or delayed surgery
  - 18 received chemotherapy
  - response rate to chemotherapy 73%
  - 7 received radiotherapy

- 5-year EFS = 58.8%, OS = 79.4%

- Outcome was strongly influenced by the feasibility of tumour complete resection

Conclusions:

- Development of a standardized approach to the diagnosis and management of PBL, and a prognostically relevant surgical staging system

- Proposal for multimodal treatment approach (conservative surgery followed by cisplatin-doxorubicin chemotherapy and postponed aggressive surgery on primary tumor and metastases)

- critical role of surgical resection
- adoption of an intensive multidisciplinary approach
- potential efficacy of chemotherapy.
- need for referral to highly experienced centres.