

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

Table 1 – Clinical characteristics of the patients series.

Clinical findings	
Age	1 month–17 years (median 4 years 9)
Gender	females – 7, males – 13
Tumour site within pancreas	head – 5, body – 4, tail – 4, body/tail – 4, head/body/tail – 3
Tumour size	<5 cm – 3, 5–10 cm – 7, >10 cm – 10
Stage	I – 6, II – 0, III – 5, IV – 9
Distant metastases	liver – 2, lungs – 2, distant lymph nodes – 2, subcutaneous tissue – 1 missing data – 2

Table 3 – Univariate analysis for 5-year event-free survival (EFS).

		EFS (%)
Tumour size	Whole series	58.8%
	<10 cm	56.0%
	>10 cm	64.8%
P (test log-rank)		0.405
Tumour location	Head of the pancreas	50.0%
	Other sites	60.5%
P (test log-rank)		0.481
Stage	Stage I	75.0%
	Stage III–IV	52.9%
	P (test log-rank)	0.390
Surgery	R0 (initial or delayed resection)	75.0%
	other	28.6%
	P (test log-rank)	0.0141

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.