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The incidence of rare, malignant or borderline tumours in children in France is difficult to evaluate at the present time. Only patients with severe, recurrent or complicated forms are referred to oncology departments. The incidence of a large number of rare diseases is therefore severely underestimated, giving a false impression of the true incidence of the disease. The *Registre National des Tumeurs Solides de l'Enfant* (RNTSE) (French Registry of Solid Tumours in Children) provides epidemiological data on childhood cancers in France. However, certain rare tumours are not included in this registry, either because their malignant nature remains uncertain or because they are managed by adult oncology teams or paediatric specialities other than oncology, less accustomed to systematic patient registration. The RNTSE also only records tumours occurring in children under the age of 15 years in metropolitan France. Concomitant registration of borderline tumours is performed but is not comprehensive. The exceptional nature of these diseases as well as their heterogeneous management in terms of both the medical specialty involved (surgery, dermatology, ophthalmology, etc.) and the site of treatment (private clinic or public hospital, paediatric or adult medicine department) account for these difficulties.

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The creation of a rare childhood tumour group by the *Société Française des Cancer de l'Enfant et de l'Adolescent* (SFCE) in November 2006 was the first step designed to address these issues. This group, mainly composed of paediatricians, surgeons, radiotherapists, and pathologists, meets at least twice a year to discuss a specific theme (aggressive vascular tumours, neuroendocrine tumours, mucoepidermoid carcinoma, etc.). Since the creation of this group, diagnostic and treatment guidelines have been proposed for certain tumours such as adrenal cortical adenomas, pleuropulmonary pneumoblastomas, pancreatoblastomas, melanomas, etc., based on retrospective analyses of patients previously treated in France, data of the literature and proposals from the other European or international rare tumour groups. These clinical practice guidelines are then made available to clinicians via the SFCE web site. A local representative responsible for rare tumours has been designated in each of the 30 SFCE centres throughout France to ensure diffusion of these guidelines. The presence of surgeons in the group also ensures close collaboration with the other members of the *Société Française de Chirurgie Pédiatrique* (SFCP). Another objective of this group is to improve the knowledge and treatment of these rare tumours by creating, in collaboration with the SFCE and the RNTSE, a national database for collection of medical information concerning these various diseases. Clinical, laboratory and radiological characteristics, the treatments administered and outcome of the disease will be recorded. Current treatment guidelines, initiated in the context of the SFCE rare tumours group since 2006, constitute the basis for setting up this database. This database therefore concerns diseases corresponding to various histologies, sharing in common their very low incidence (less than

D.T. Schneider et al. (eds.), *Rare Tumors In Children and Adolescents*, Pediatric Oncology,
DOI 10.1007/978-3-642-04197-6_7, © Springer-Verlag Berlin Heidelberg 2012

2 cases per million children under the age of 18) and the absence of formal treatment guidelines. For practical purposes, tumours with an exceptionally low incidence, but for which treatment guidelines or data collection are already available in the context of the SFCE or SIOP working party, will not be included in this group. It was also arbitrarily decided not to include in this rare childhood tumour group those rare haematological malignancies included in the 'leukaemia group'. Consequently, the main diseases concerned by this group are: undifferentiated nasopharyngeal carcinoma; pancreatoblastoma; Frantz's tumour (pseudopapillary tumour of the pancreas); pleuropulmonary blastoma; pseudo-inflammatory tumour; mesothelioma; thymoma; gastrointestinal stromal tumours; desmoplastic small-cell tumour; adrenal cortical adenoma; malignant phaeochromocytoma; carcinoid of the appendix; carcinoid of the small intestine; carcinoid tumour of the bronchus; midline carcinoma; aggressive giant-cell bone tumours; chondroblastoma; chondrosarcoma; malignant head and neck tumours: sialoblastoma, mucoepidermoid carcinoma, aggressive benign vascular tumour, lung carcinomas, urothelial carcinomas and chordomas. Some tumours that are common in adults but rare in children will also be included in this database when they occur in children, such as cutaneous or choroidal malignant melanomas, ocular medulloepitheliomas, breast cancers, colon cancers or thyroid cancers.