

## Defining and listing very rare cancers of paediatric age: consensus of the Joint Action on Rare Cancers in cooperation with the European Cooperative Study Group for Pediatric Rare Tumors

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According to the consensus, very rare paediatric cancers were identified as those with an **annual incidence <2/1000000** and corresponded to 11% of all cancers in patients aged 0-14 years.

Two subgroups were identified: tumour types typical of childhood (i.e. hepatoblastoma, pleuropulmonary blastoma, pancreatoblastoma) and those typical of adult age (i.e. carcinomas, melanoma).

The threshold of 2/1000000 could also be adopted in populations aged 0-19 years: in this case, three tumour types had an incidence rate which was >2/1000000 (i.e. thyroid and testicular cancers and skin melanoma), but the consensus experts considered them as 'very rare' according to their clinical needs (e.g. shortage of knowledge and clinical expertise as the other rare paediatric cancers).

*To establish a shared definition and produce a list of these entities, the European Union Joint Action on Rare Cancers (JARC) promoted a consensus effort. The definition was based on the incidence rates estimated using the information network on rare cancers (RARECAREnet) database, pooling data from 94 population-based cancer registries and 27 countries.*

*The RARECAREnet list of cancers was used to estimate the incidence rates.*

*This list groups cancers by combining the International Classification of Diseases for Oncology, third edition, morphology and topography codes.*

The JARC/EXPeRT consensus produced a definition and a list of very rare paediatric cancers (founded on the incidence rate of these malignancies in Europe), which may represent a starting point for prioritising research on these tumours, based on data and patients' clinical needs.

This effort also wants to differentiate very rare tumours from other paediatric cancers and might help in suggesting dedicated methodological approaches for research.

Noteworthy, the list should be used flexibly and seen as a 'work in progress' because new very rare tumours are coming to light every year, as we learn more about the molecular basis of many cancer types.