Guidelines to be followed by centres, services and units in order to be designated as Reference Centres, Services and Units of the National Health System as agreed by the Interterritorial Board.

5. EXTRAOCULAR TUMOURS IN CHILDHOOD (Rhabdomyosarcoma)[1,2,3]

The *rhabdomyosarcoma* is the most common soft tissue sarcoma in children. The orbital rhabdomyosarcoma, malignant tumour arising from the muscle striations of the oculomotor muscles, is the most frequent malignant tumour in childhood.

Multidisciplinary approach: medical and radiotherapy oncology and ophthalmology (with paediatric and orbital ophthalmology units). It is also required to have an anatomical pathology team with experience in sarcomas, since the histological characteristics are decisive in the treatment and diagnosis.

Surgery, chemotherapy and radiotherapy are used for *treatment*. Diagnosis depends on tumour site, size, resectability, presence of metastasis and the histological type. Global surviving after 5 years is higher than 70%[1,2].
Although the main treatment is chemotherapy and radiotherapy, it is beneficial to have a surgeon with experience in orbital surgery since biopsies are required for diagnosis. In some cases, excisional biopsy is performed according to protocols. After recurrence radical orbital surgery is indicated in some cases. The orbital surgery approach to the intraconal and extraconal spaces is less traumatic and leaves minimal scar, which is important given that in recurrence cases chemotherapy is less effective in tissues with surgical scars.

It is difficult to assess the effectiveness of the current protocols used in the treatment of extraocular tumours in childhood in terms of mortality and morbidity, given its rarity, hence the need to bring them together in reference units.

A. *Rationale for the proposal*

► Epidemiological data on extraocular tumours in childhood (incidence and prevalence).	*Incidence*[1,2,3]*:* There is a long list of orbital tumours, all with low incidence. Rhabdomyosarcoma represents 3-4% of all malignant tumours in children under 15. Its annual incidence in Spain is unknown; however it is supposed to be similar to the incidence in the USA, 4 to 7 cases out of every million of white children under 15. In 40% of the cases the tumour appears either in head or neck, and in 10-20% of the cases it is located in the orbit.

B. ***Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units treating extraocular tumours in childhood.***

► Experience of the Reference Centres, Services and Units: - Activity: • Number of procedures on extraocular tumours in childhood that should be performed in a year to ensure an adequate care	 - At least an average of one new patient with rhabdomyosarcoma every year in the last 5 years. - 20 surgeries of orbital tumours in a year.
- Other data: research on the subject, postgraduate teaching, continuing training, etc.	- Accredited postgraduate teaching. - Participation in research projects and publications in the field[a]. - Continuing training programs[a].
► Specific resources of the Reference Centres, Services and Units: - Human resources required for the adequate care of extraocular tumours in childhood. Professional experience[b]**.**	Existence of a Hospital Committee on tumours with an updated action protocol based on scientific evidence. - Multidisciplinary team: Ophthalmologist, medical oncologist, and radiotherapeutic oncologist. - 24 hour continuous ophthalmic care. - Nursing staff, surgical auxiliaries and technicians. Multidisciplinary team with at least 2 year experience in extraocular tumours in childhood: - Ophthalmologists with experience in paediatric patients and orbital surgery. - Medical oncologist with experience in paediatric patients and in the treatment of rhabdomyosarcoma in the eye and other sites. - Radiotherapeutic oncologist with experience in paediatric patients and ocular pathology.

- Specific equipment required for the adequate care of extraocular tumours in childhood.	- Nursing staff with experience in paediatric patients and ocular pathology. - Specific equipment required for orbital surgery: • Palpebral surgery standard equipment. • Malleable separators (used in neurosurgery). • Micromotor system. • Oscillating saw, for orbitotomy with osteotomy. • Biological glue and bone wax. • Frontal lighting source.
► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of extraocular tumours in childhood.	- Radiotheraypy, including brachytherapy. - Anaesthesia services with experience in paediatric patients[b]. - Intensive care unit with experience in paediatric patients[b]. - Paediatric services. - Neurosurgery services with experience in paediatric patients[b]. - Neurophysiology. - Image diagnosis: CT scan, MRI, ultrasound, nuclear medicine. - Anatomical pathology services with experience in sarcomas[b]. - Haematology services.
► Procedure and clinical results indicators of the Reference Centres, Services and Units [c].	**The indicators will be agreed with the Units that will be designated.**
► Existence of an adequate IT system (Type of data that the IT system must include to allow identification of the activity and evaluation of the quality of the services provided)	- Filling up the complete MBDS of hospital discharge. - The unit must have a *registry of patients* with extraocular tumours which at least must include: - Data required for the tumour registry of the hospital: patient's identity (medical record number), date of birth, sex, address, tumour site (International Classification of Diseases for Oncology), date of diagnosis and recurrence, diagnosis method, tumour histology, stage. Initial treatment. If applicable, cause and date of death.

	- Admission date and discharge date. - Date of surgery. - Diagnosis procedures performed to the patient (ICD-9-CM). - Main diagnosis (ICD-9-CM). • Injury etiology. • Injury characteristics, size and site. - Number and type of therapeutic procedures provided to the patient (ICD-9-CM): • Surgical procedures (orbital surgery). • Other therapeutic procedures. - Date when the different procedures were performed. - Treatment results. • Local recurrence. - Complications and solutions (ICD-9-CM). - The unit must have the required data which should be sent to the Spanish National Health Service Reference Centres, Services and Units Appointment Commission Secretariat for yearly reference unit monitoring.

[a] *Criteria to be assessed by the Appointment Commission.*
[b] *Experience will be accredited by certification from the hospital manager.*
[c] *Clinical results standards, agreed to by the experts group, will be assessed, initially by the Appointment Commission, while in the qualification process, as more information from the Reference Centres, Services and Units is being obtained. Once qualified by the Appointment Commission, the Quality Agency will authorize its compliance, as for the rest of guidelines.*

Bibliography:

[1] Diseases of the orbit. Jack Rootman. Lippincott Company. 2ª edición. 2003.
[2] Patología orbitaria. Pérez Moreiras JV. Edika Med. 2000.
[3] Rabdomiosarcoma orbitario: Dificultades con el protocolo de tratamiento europeo. Arch Soc Esp Oftalmol 2005; 80: 331-338.