

***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.***

## **6. INTRAOCULAR TUMOURS IN CHILDHOOD (Retinoblastoma)<sup>1,2,3,4,5</sup>**

From the intraocular tumours, the *retinoblastoma* is the most frequent malignant intraocular tumour in childhood; it is a malignant tumour in the retina.

The *diagnosis* requires ophthalmological and biomicroscopic examination under general anaesthesia, ultrasound scan to measure the tumour, and CT scan and MRI to rule out the spread of the tumour.

There are different *types of treatment* according to size, site or evolution stage: chemoreduction, photocoagulation, cryocoagulation, transpupillary thermotherapy, external radiotherapy and brachytherapy, enucleation and exenteration.

Therefore, the approach to intraocular tumours in childhood is multidisciplinary: medical and radiation oncology, and an ophthalmologist with experience in retinal surgery.

### ***A. Rationale for the proposal***

► Epidemiological data on intraocular tumours in childhood (incidence and prevalence).	Incidence of 1 out of every 15,000 live births (approximately 35 per year).
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### ***B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units treating intraocular tumours in childhood.***

► Experience of the Reference Centres, Services and Units:	
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<p>- Activity:</p> <ul style="list-style-type: none"> <li>• Number of cases of intraocular tumour in childhood that should be assisted in a year to ensure an adequate care.</li> </ul> <p>- Other data: research on the subject, postgraduate teaching, continuing training, etc.</p>	<p>At least 6 new patients with intraocular tumours per year.</p> <ul style="list-style-type: none"> <li>- Accredited postgraduate teaching.</li> <li>- Participation in research projects and publications in the field<sup>a</sup>.</li> <li>- Continuing training programs<sup>a</sup>.</li> </ul>
<p>► Specific resources of the Reference Centres, Services and Units:</p> <p>- Human resources required for the adequate care of intraocular tumours in childhood.</p> <p>Professional experience<sup>b</sup>:</p> <p>- Equipment required for the adequate care of intraocular tumours in childhood.</p>	<p>Existence of a Hospital tumour board with an updated action protocol based on scientific evidence.</p> <p>Multidisciplinary team: Ophthalmologist, medical oncologist, and radiation oncologist.</p> <ul style="list-style-type: none"> <li>- 24 hour continuous ophthalmic care.</li> <li>- Nursing staff, surgical auxiliaries and technicians.</li> </ul> <p>Multidisciplinary team with at least two year experience in intraocular tumours in childhood:</p> <ul style="list-style-type: none"> <li>- Ophthalmologists with experience in paediatric patients and retinal surgery.</li> <li>- Medical oncologists and radiation oncologists with experience in paediatric patients and ophthalmologic pathology.</li> <li>- Nursing staff with experience in paediatric patients and ocular pathology.</li> </ul> <ul style="list-style-type: none"> <li>- Ophthalmological and biomicroscopic examination.</li> <li>- Availability of the different types of possible treatments for intraocular tumours in childhood according to size, site or evolution stage: chemoreduction, photocoagulation, cryocoagulation, transpupillary thermotherapy, enucleation and exenteration.</li> <li>- Radiotherapy, including brachytherapy</li> </ul>

<p>► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of intraocular tumours in childhood.</p>	<ul style="list-style-type: none"> <li>- Anaesthesia services with experience in paediatric patients<sup>b</sup>.</li> <li>- Intensive care unit with experience in paediatric patients<sup>b</sup>.</li> <li>- Paediatric Services.</li> <li>- Neurosurgery services with experience in paediatric patients<sup>b</sup>.</li> <li>- Image diagnosis: CT scan, MRI, ultrasound, nuclear medicine.</li> <li>- Neurophysiology.</li> <li>- Anatomical pathology services with experience in this type of tumours<sup>b</sup>.</li> <li>- Haematology services.</li> <li>- Genetics Unit.</li> </ul>
<p>► Procedure and clinical results indicators of the Reference Centres, Services and Units <sup>c</sup>:</p>	<p><b>The indicators will be agreed with the Units that will be designated.</b></p>
<p>► 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)</p>	<ul style="list-style-type: none"> <li>- Filling up the complete MBDS of hospital discharge.</li> <li>- The unit must have a <i>registry of patients</i> with intraocular tumours which at least must include: <ul style="list-style-type: none"> <li>- 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.</li> <li>- Admission date and discharge date.</li> <li>- Date of surgery.</li> <li>- Diagnosis procedures performed to the patient (ICD-9-CM). <ul style="list-style-type: none"> <li>• Ophthalmological and biomicroscopic examination under general anaesthesia.</li> <li>• Ultrasound, CT scan and MRI.</li> </ul> </li> <li>- Main diagnosis (ICD-9-CM). <ul style="list-style-type: none"> <li>• Injury etiology.</li> <li>• Injury characteristics, size and site.</li> </ul> </li> <li>- Number and type of therapeutic procedures provided to the patient (ICD-9-CM): <ul style="list-style-type: none"> <li>• Chemoreduction, photocoagulation, cryocoagulation, transpupillary thermotherapy,</li> </ul> </li> </ul> </li> </ul>

	<p>external radiotherapy and brachytherapy, enucleation and exenteration.</p> <ul style="list-style-type: none"> <li>• Other therapeutic procedures.</li> </ul> <p>- Complications (ICD-9-CM).</p> <p>- 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.</p>
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<sup>a</sup> *Criteria to be assessed by the Appointment Commission.*

<sup>b</sup> *Experience will be accredited by certification from the hospital manager.*

<sup>c</sup> *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:**

<sup>1</sup> Tumores intraoculares. Encinas JL. Ed. Univ Ramón Areces, year 2006.

<sup>2</sup> Actualización en tumores intraoculares. Capeans Tomé C, Mesa Redonda 75th Congress of the Spanish society of ophthalmology, 1999.

<sup>3</sup> Ophthalmic Pathology and Intraocular Tumors. Basic and Clinical Science Course. American Academy of Ophthalmology, 2003-2004.

<sup>4</sup> Atlas of Intraocular Tumors. Sanborn, Gonder, Shields. 1994.

<sup>5</sup> Actualización en cirugía oftálmica pediátrica. Fonseca, Abelairas, Rodríguez Peralta. Ponencia SEO 2000.