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**Learning objectives**

The aim of this report is to illustrate the correlation between the notochordal function, development and anatomical path in the base of the skull regarding to location and nature of derivated lesions from this structure.

Describe benign notochordal lesion, focusing in Ecchordosis Physaliforma and its implication in the differential diagnosis with intradural location of craniocervical junction chordomas.

Review of chordomas of the craniocervical junction and their imaging findings. Exposure of the main features of other diseases included in their differential diagnosis.

**Background**

**Notochorda development**

Notochord formation begins between the third and fourth week of embryonic development. It is originated from invaginated cells into the primitive streak which are arranged with cranial direction forming a median cellular cord, the notochordal process. When this process acquires a lumen, it becomes in the notochordal canal.

The notochordal process is placed between the endoderm and ectoderm and extends into the cranial region until it reaches the prechordal plate, where the ectoderm and endoderm are consolidated. Endoderm of the oropharyngeal membrane arises in prechordal plate, representing the location of future oral cavity. The notochordal process concludes with proliferation of notochordal plate cells, which are removed from endoderm to form the definitive notochord.

The remaining embryonic tissue around notochord forms the vertebrae and notochord persists as the central part of the vertebral body. The notochord undergoes a process of regression during fetal life, except in place of future intervertebral discs which contribute to the formation of the nucleus pulposus.
Notochord ascends between body vertebraes in its axial course until leaving the axis. Notochord gets into the basiocciput with a curved path and finalizes in the body of the sphenoid, just caudal to the pituitary fossa, at the dural margin, where the cephalic end of notochord is found. Fig. 1 on page 11

Fig. 1: Scheme of a sagittal view of the skull base exemplifying the developmental anatomy of the notochord. The cephalic end of the notochord is entrapped in the basiocciput and posterior sphenoid.

References: Hospital universitario La princesa - Madrid/ES

The notochord is the central axle to the formation of the axial skeleton and has an inductor role in development of central and peripheral nervous system.

In addition, the origin of the posterior cranial base is directly related to the notochord.

In the vicinity of cephalic end of notohord, chondroid nucleus of ossification are formed, which will lead to different bone structures of the skull base.

These chondroid nucleus of ossification include, among others, prechordal cartilages (located anterior regarding to notochord) which are precursors of the sella and the posterior body of the sphenoid, and parachordal cartilages or basal lamina (located around the notochord) which are the precursors of basioccipital bone.
Some studies suggest that the notochord could play an organizing role through inducing the chondroid differentiation and segmentation of the mesenchymal elements of the vertebral bodies, which may explain the variant of chondroid chordoma.

**Entities related to notochord**

It is postulated that the process of regression and atrophy occurring in the notochord is not an uniformed process. Cephalic and caudal ends of notochord are not only confined to the notochord body but are composed of bifurcations and separate fragments of notochord body, explaining the possibility of finding notochordal cells in those locations. This theory about anatomical path of the notochord in the skull base during embryological development, could account for the different locations that entities related to the notochord acquired:

**Fig. 2 on page 12**

- Anatomic variations which are considered normal remnants of the notochord (fossa navicularis, canalis basilaris medianus)

- Disorders whose origin is closely related notochord (Tornwaldt's cyst).

- Benign lesions derived from the notochord (Ecchordosis Physalifora of retroclival location).

- Malignant lesions arising from the notochord (extraosseous and intraosseous chordomas).
Fig. 2: Representative diagram about entities related to notochord and correlation between their location and notochordal path in skull base.

References: Hospital universitario La princesa - Madrid/ES

At the point where the anterior clivus path of the notochord and primitive endoderm pharyngeal get in contact, forces of regression and proliferation occur, forming the nasopharyngeal bursa. Some situations like inflammatory process or adenoidectomy can occlude the nasopharyngeal bursa, forming the Tornwaldt's cyst.

Tornwaldt's cyst is easily recognizable on MRI. It is situated in the midline and shows a marked hyperintensity in T2-weighted images, as well as in T1-weighted images due to proteinaceous content. It is an incidental finding in most of situations and does not require injection of gadolinium. If contrast administration is performed Tornwaldt's cyst demonstrates absence of enhancement related with its mucosa origin. This condition frequently coexists with the fossa navicularis, a small osseous recess on anterior side of the basilar apophysis of the occipital bone representing the point where notochord leaves basisphenoid in its embryonic development. Fig. 3 on page 12 Fig. 4 on page 13

There are other bone defects that are normal variants and are also related to the embryonic notochord path passing through the basiocciput, as the canalis basilaris medianus. It is postulated as a vestige of the notochordal canal (canalis chordae) in the postnatal life. Knowledge about these anatomical variations allows not confuse them with fracture lines.
Ecchordosis physaliphora

Ecchordosis Physalifora (EP) is a benign lesion, often incidental in MRI studies, with low incidence, found in approximately 2% of autopsies. EP is considered as a small, gelatinous, hamartomatous mass located on intradural space between protuberance and clivus.

Initially Luschka and Virchow in 1857 coined the term "physaliphora ecchondrosis" believing that this structure was derived from a degenerative process of the sphenoorbital synchondrosis. Müller recognized in 1858 its notochordal origin and in 1894, Rippert called it Ecchordosis Physalifora.

This entity is typically described in the intradural space in retroclival location, although it has also been observed in the dens of the axis and the coccyx.

Histologic features of EP are the presence of physaliferous cells, which typically contain large vaculolas inside. These cells are also common in chordomas, and immunohistologically are distinguished by immunoreactivity to cytokeratins.

EP has received sundry nomenclatures (giant notochordal rest, and giant notochordal hamartoma of intraosseous origin, etc.), and typically has been considered as a remnant of the notochord, although controversy exists today. Several reports have remarked that fetal remains of the notochord not show immunoreactivity for cytokeratin 18, whereas physaliferous cells are positive to them.

In addition, a sclerotic reaction in adjacent bone has been reported, so EP would not be considered as an authentic notochordal vestige. Therefore, some authors propose to include this entity in cell tumor benign notochordal, typically retroclival referring to EP.

The differential diagnosis between EP and chordomas of intradural location is often complicated, as they share the same location and histopathologic characteristics because both are neoplasms derived from the notochord, while the EP has a benign character and chordomas malignant nature. However, they have distinctive features: Table 1 on page 14.
• EP is usually asymptomatic due to their small size and indolent growth patterns, unlike chordomas.

• EP shows no enhancement in contrast-enhanced sequence of MRI.

• Inconspicuous sclerotic reaction on adjacent bony structures.

• Histomorphologically, EP has osseous trabeculae preserved, polygonal and large vacuolated cells similar to adipose cells, and absence of nuclear atypia, mitosis, necrosis or extracellular myxoid matrix, compared to chordomas.

• Age of appearance: younger in EP, older in chordomas.

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**Table 1:** Descriptive table of the most important features in the differential diagnosis between Ecchordosis Physaliforma and intradural extraosseous chordomas.

**References:** Hospital universitario La princesa - Madrid/ES

The distinction between EP and chordoma is extremely important due to the prognosis implications and therapeutic approaches. Chordomas are characterized by an invasive nature and high rate of recurrence, which confines them to a worse prognosis, while the EP may be followed with observation although there is no consensus on treatment.

**Chordomas of craniocervical junction**
Chordomas are a rare bone malignant neoplasm that are thought to arise from transformed remnants of notochordal tissue, a primitive cell line around which the skull base and the vertebral column develop, as previously discussed. The overall incidence of chordomas is 0.08-0.5 per 100,000. These tumors affect more men than women and have a peak incidence between 50-60 years of age, with a very low incidence in patients younger than 40 years. Chordoma is uncommon in children, but when present, it is more aggressive. Rarely, chordoma can be familial.

Although the sacrum has been the most frequent location, actually evidence suggests an almost equal distribution in the skull base (32%), spine (32.8%), and sacrum (29.2%). In this review, we will focus on intracranial chordomas. They account for 1% of intracranial tumors and 4% of all primary bone cancers.

Most intracranial chordomas are intraosseous, although there have been some cases of extraosseous chordomas, located in the dural layer (intradural chordomas) and nasopharynx (nasopharyngeal chordomas). Recent advances in immunohistochemical studies and the discovery of a gene duplication in the transcription factor T gene (brachyury) have allowed us to distinguish these extraosseous tumors of other pathology in the same location.

Histologically, chordomas exhibit physaliferous cells as typical pathologic feature (but not pathognomonic) and immunoreactivity for S-100 and epithelial markers such as epithelial membrane antigen (MUC1) and cytokeratins. Chordomas can be divided into three histological types according to the degree of histologic atypia that exhibit:

1) Classical chordoma: Physaliferous cells are arranged in cords set in a pale matrix of mucopolysaccharide and separated by fibrous septa. Areas of necrosis, recent and old hemorrhage, and entrapped bone trabeculae are seen.

2) Chondroid chordoma: their stroma resembles hyaline cartilage with neoplastic cells in lacunae, which is histologically similar to low-grade chondrosarcomas. Pathologic differential diagnosis between them was difficult because of their shared S-100 immunoreactivity. But brachyury expression by cells chordoma, as recent molecular finding, combined with cytokeratin staining has enabled differentiating between chondroid chordoma and chondrosarcoma with 98% sensitivity and 100% specificity.

3) Dedifferentiated chordoma: extremely rare, highly aggressive and usually found in the sacrococcygeal region.
Intracranial chordomas of intraosseous location originate mainly in the sphenoo-occipital synchondrosis of the clivus. Others sites of origin include the petrous apex, the sellar area, sphenoid sinus, maxilla and paranasal sinuses.

From the clivus chordoma can extend: Fig. 5 on page 14

- Anteriorly: sellar and parasellar areas, sphenoid sinus, posterior ethmoid sinus.
- Laterally: petrous apex, middle cranial fossa.
- Posteriorly: prepontine cistern, brainstem (mainly pons).
- Inferiorly: foramen magnum, jugular fossa, atlas and other cervical vertebrae, nasopharynx, parapharyngeal space.
- Superiorly: optic chiasm, third ventricle.

In addition, intracranial chordomas commonly encroach on the anterior visual pathway and on the cranial nerves in the prepontine cistern and cavernous sinus, resulting in visual and cranial nerve abnormalities.

Chordomas are considered as slow-growing and locally invasive tumors. Because of this they produce symptoms insidiously, which vary with tumor location and proximity to critical structures. Diplopia (cranial nerve palsy) and headache (occipital or orbitofrontal) is the most common initial complaint. Large tumors can also involve function of lower cranial nerves.

Despite the improvement in surgical and radiotherapy treatment, the management of skull base chordomas remains a challenge. They are generally slow-growing tumours that are locally aggressive and invasive, and spread along bone and neurovascular structures makes management of these patients difficult. An optimum treatment paradigm is maximally safe cytoreductive surgery and advanced radiation delivery technique, with an enhanced emphasis on preservation of neurological functions. Therefore, it is not uncommon the recurrence and progression of these tumors. But they rarely metastasize.

**Differential diagnosis of craniocervical junction chordomas**
Chondrosarcomas and skull base chordomas have led to confusion in the literature, due to they share similar intracranial location, clinical presentation and radiologic characteristics, especially chondroid chordoma variant, which also presents cartilaginous elements so differential diagnosis between both often is extremely complicated.

Chondrosarcomas derive from primitive mesenchymal cells or from the embryonic rest of the cartilaginous matrix of the cranium. They are present most frequently in the long bones and pelvis and less of 10% have been observed in head and neck. Location of chondrosarcomas and chordomas and their growth pattern appear to be also different, with regard to chondrosarcomas would begin laterally and would grow into the midline, unlike chordomas with midline origin and growth laterally. Immunohistologically techniques help in differential diagnosis given that chordomas, chondroide variant included, are positive to cytokeratins and epithelial membrane antigen (EMA), whereas chondrosarcoma are negative for these markers.

Intracranial chondrosarcomas often are of the classic subtype and low-grade, so they have better prognosis than chordomas, with less disease recurrence and minor risk of metastases, which allow control them satisfactorily by surgical resection. Chondrosarcomas may benefit from adjuvant radiotherapy although the risk-benefit is not established regarding to surgical treatment which achieved control of these tumors. However, the optimal treatment of chordomas (including chondroide variant) incorporates surgical resection associated with high-dose radiotherapy due to their high rate of disease recurrence and aggressive clinical course.

Cavernous hemangiomas are common vascular malformations which have a strong female predominance and more incidence in the middle age. When esphenoidal bone involvement is present they have to be included in differential diagnosis with chordomas. The cavernous hemangioma may present erosion of adjacent bony structures. Hemangiomas displace but not infiltrate surrounding vascular structures, and can give clinical symptoms derivated of compression related with large size. Its imaging behavior makes possible diagnosis because although they are hyperintense in T2-weighted MRI images like chordomas, the typical feature of hemangiomas is an heterogeneous centripetally "filling-in" enhancement pattern.

Meningiomas are the most common benign intracranial tumors, representing 13%-19% of all intracranial neoplasms. They arise from arachnoid layer cells and appear where meninges are observed. Clival meningiomas show a characteristic dural attachment and do not produce lytic bone lesion. Instead, they cause bone sclerosis and manifest homogeneous enhancement.
Plasmocytoma and lymphoma occasionally present osseus affectation in the skull base producing lytic bone destruction when are located in the clivus. If they acquire a central location these tumors can mimic skull base chordomas.

Craniopharyngiomas often demonstrate typical imaging features and are located more anteriorly and superiorly in midline respecting intracranial chordomas.

Skull base metastases in absence of primary tumor have been poorly described.

Rhabdomyosarcoma should be considered in pediatric patients. It usually begins in the nasopharynx and shows as a large mass with associated lytic bone destruction.

Other differential diagnoses with intracranial chordomas include aggressive pituitary adenoma, histiocytosis X, dermoid and epidermoid cysts, trigeminal neuroma, and fibrous dysplasia.

Images for this section:
Fig. 1: Scheme of a sagittal view of the skull base exemplifying the developmental anatomy of the notochord. The cephalic end of the notochord is entrapped in the basiocciput and posterior sphenoid.

Fig. 2: Representative diagram about entities related to notochord and correlation between their location and notochordal path in skull base.
Fig. 3: Tornwaldt's cyst associated with both fossa navicularis and canalis basilaris medianus. A. Sagittal CT demonstrates a defect on the intracranial surface of the basiocciput, like a well-defined channel in the midline, suggested of canalis basilaris medianus (green arrow). B. Axial CT shows notchlike defect in the basiocciput suggested of fossa navicularis (yellow arrow), often associated with Tornwaldt's cyst. C. Axial FIESTA MR image obtained through the nasopharynx shows a high-intensity cyst (red arrow) in the expected location of the pharyngeal bursa of Luschka. This Tornwaldt's cyst was fortuitously discovered and asymptomatic. D. Sagittal T1-weighted gadolinium-enhanced MR image reveals absence of enhancement of the cyst (pink arrow).
Fig. 4: Note osseus notch (yellow arrows) on the anterior surface of the clivus regarding fossa navicularis.

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Table 1: Descriptive table of the most important features in the differential diagnosis between Ecchordosis Physalifora and intradural extraosseous chordomas.
**Fig. 5:** Tumor spread. Sagittal (A) and coronal (B) contrast-enhanced T1-weighted MR images show a lesion arising from the skull base. Laterally, the mass involves both cavernous sinuses, with more involvement on the right side with displacement and narrowing of the intrapetrous carotid artery (red arrow). Anteriorly, the lesion enlarges the sella turcica and reaches the sphenoid sinus (green arrow). Posteriorly, we identify two nodular projections of the mass (blue arrows) encasing and narrowing basilar artery (yellow head arrow). Brainstem is deformed. Inferiorly, the mass extends into the odontoid process. Superiorly, the lesion involves suprasellar cistern and anterior recesses of the third ventricle (orange arrow).
Findings and procedure details

Imaging findings of Ecchordosis Physalifora

Intracranial EP is located typically in intradural space of prepontine cistern region, behind of clivus. A stalk connecting this lesion with the dorsal wall of the clivus has been suggested as a hallmark of EP. Characteristically EP appears hypointense on T1-weighted images and hyperintense on T2-weighted images, without contrast enhancement, issue that allows differentiation regarding to intradural chordomas. Fig. 6 on page 20

Imaging findings of skull base chordomas

Diagnostic imaging of skull base chordomas is mainly focused on CT and MRI. Both techniques define more accurately diagnostic characteristics and tumor extension, that is crucial information both the surgeon and the radiation oncologist for better treatment and improved prognosis of these patients.

CT is very accurate in the depiction of bone abnormalities. We can improve sensitivity on the detection of lesions of the skull base, if we use a bone and soft-tissue algorithm. Study of skull base should include thin-section axial and coronal unenhanced and contrast material-enhanced images. However, keep in mind that the capacity of CT to show soft-tissue structures in the posterior fossa is limited due to beam-hardening artifacts.

MRI is considered the single best modality for radiologic evaluation of intracranial chordomas. MR imaging is similar of CT in detecting these tumors but is considerably superior to CT in the delineation of the lesions with respect to volume and relation to adjacent neural structures. But it is limited only in the evaluation of calcification and cortical bone. MR study includes multiplanar sequences. Sagittal images are the most valuable for defining the posterior margin of the tumor. In this plane we can show the relation between the tumor and brainstem, depict nasopharyngeal extension of the tumor and disclose transdural transgression by a tumor. Coronal images show extension of the tumor into the cavernous sinuses and depict the position of the optic chiasm and tract.

On CT, the skull base chordoma is typically a midline located, well-circumscribed, hyperdense mass with lytic changes of the clivus. Common CT features of chordoma
include bone destruction, intratumoral soft tissue component, and a sharp margin separating the tumor from adjacent normal soft tissue or bone. Fig. 7 on page 20

Intratumoral densities are frequently seen on CT and are thought to represent sequestra from bone destruction rather than true new bone formation (dystrophic calcifications). The chondroid variant is more likely to demonstrate real intratumoral calcifications. Unlike chondrosarcomas, chordomas do not form calcified rings or arcs. Extension into posterior fossa, sella, sphenoid sinus, as well as cavernous sinus is common Fig. 8 on page 21

Intracranial chordomas show moderate to high enhancement on CT.

On MRI, the bulk of the tumor is isointense or hypointense on T1-weighted images and is easily recognized within the high signal intensity of the clival fat (Fig. 9 on page 22). Localized areas of intratumoral hyperintensity represent hemorrhage or mucoid materials (Fig. 10 on page 23). We can confirm the presence of hemorrhagic foci with gradient-echo imaging that is susceptible to blood, at which the foci appear as dark areas.

The classic appearance of intracranial chordomas at T2-weighted images is high signal hyperintense secondary to high fluid content or abundance of physaliferous cells. Fig. 11 on page 24

Intratumoral areas of T2 hypointensity are secondary to chronic hemorrhage, bone fragments, or highly proteinaceous mucoid material. Low-signal-intensity septations that separate high-signal-intensity lobules are commonly seen and give the gross appearance of a dirty cauliflower (multilobulated mass) Fig. 12 on page 25

The condroid variant may be less bright as typical chordoma on T2-weighted MR images due to a watery gelatinous matrix is replaced by cartilaginous foci.

Usually chordomas enhance intensely with contrast (Fig. 13 on page 26), unless the tumor is completely necrotic or contains a large amount of mucinous material in the tumor ("honeycomb" appearance). In these cases, the enhancement is slight or even absent (Fig. 14 on page 26). Fat suppression is useful for differentiating enhanced tumor margins from adjacent bright fatty bone marrow.

MR angiography allows better evaluation of displacement or encasement of intracranial arteries by the tumor, being visualized in up to 79% of intracranial chordomas. Despite the high frequency of intracranial arterial involvement, arterial narrowing is rare in intracranial chordomas. Cerebral angiography has been displaced by MR angiography and is reserved for cases in which there is significant displacement, encasement or narrowing
of the internal carotid or vertebral artery at MR angiography. Cerebral angiography can better demonstrate the degree of luminal narrowing or occlusion and the extent of collateral circulation (Fig. 15 on page 27) Venous involvement or occlusion is also readily visualized at MR venography.

**Imaging findings in differential diagnosis of skull base chordomas**

Chordomas typically show hyperintense signal on T2-weighted images, unlike other tumor pathologies, so the main differential diagnosis with chordomas skull base should encompass T2 hyperintense masses which share same location. Some of these are briefly exposed below:

*Chondrosarcomas* represent the main differential diagnosis of chordoma due to similar clinical presentation, histological and imaging characteristics. From the imaging perspective, chondrosarcomas are akin with respect to chordomas; hypo-isointense on T1-weighted images, hyperintense on T2-weighted images, and assidously enhanced with gadolinium. However, location can give some clue in their distinction cause condromas occupy more frequently the petroclival fissure, unlike chordomas which tend to lie anterolateral from midline region. Fig. 16 on page 28

*Cavernous hemangiomas* are well-define masses in parasellar region, strightlly hyperdense and homogeneous enhacement after administration of material contrast. The MRI findings consist of marked T2 hyperintensity, and characteristic centripetal enhancement pattern on sequential gadolinium enhanced images. Fig. 17 on page 29

*Skull base neurinomas* are infrequent, and most often involve V cranial nerve and its branches. When VI and IV cranial nerves are implicated, neurinomas can mimic radiological findings of chordomas due to the similar location. Neurinomas are well defined lesions and they show isointensity on T1 and hyperintensity on T2- weighted images, with homogeneous enhancement after injection of gadolinium. The midline location and destructive pattern of chordomas may help in their distinction.

*Metastases* with hyperintense behaviour in T2-weighted sequence can mimic chordomas, but in absence of primary neoplasm are uncommonly in skull base of cranium. Fig. 18 on page 30
Other possible entities representing the differential diagnosis of chordomas of the craniocervical junction are:

**Myeloma affectation and plasmacytoma:** Radiological features of plasmacytoma involving the skull base are not specific. It is manifested on MRI as a lytic lesion isointense on T1-weighted images, with slightly increased signal in T2-weighted images and homogeneous enhancement after injection of gadolinium. Differential diagnosis must be considered when they are located in the clivus and sellar region. Fig. 19 on page 31

**Spheno-orbital or petroclival meningiomas** may have similar radiological findings to chordomas. CT images show them as a well-defined hyperdense mass, and T1 and T2-weighted MR images demonstrate a isointense lesion, which is characterized by a homogeneously enhancing in postcontrast images. Besides a typical dural tail is observed and they frequently associated hyperostosis, unlike chordomas, although an osseus lytic component can rarely be seen indicating malignancy and aggressiveness. Fig. 20 on page 32

**Macroadenomas** occupy and expand sella, and they can occasionally manifest lytic change of the sphenoid bone or clivus through infrasellar extension. Macroadenomas have variable imaging features, tending to appear isointense compared with gray matter on T1-weighted images, an isointense on T2-weighted images, however this feature can change due to presence of cystic or hemorrhagic areas. Contrast-enhanced MRI sequence demonstrates a large homogeneously enhancing of macroadenomas. Fig. 21 on page 33

**Fibrous dysplasia** is an idiopathic bone disease. The bone marrow is replaced by disorganized fibro-osseous tissue. The medullary cavity expands but remains intact cortical, unlike chordoma. Fibrous dysplasia has an appearance on "ground glass" is hypointense on T1, isointense on T2 and does not exhibit enhancement after gadolinium administration. In MRI can be confused with a mass lesion; the CT establishes the diagnosis. Fig. 22 on page 34

**Craniopharyngiomas** are benign but aggressive neoplasms, which arise from the Rathke pouch epithelium observed along the path of the craniopharyngeal duct. They are most commonly located in suprasellar area but often have sellar extension and can be purely infrasellar. In these cases although they are uncommon could mimic chordomas
due to presence of clival destruction, and cyst formation and contrast enhancement is rare. However, in our institution a case of craniopharyngioma with cyst formation and enhancement was described. Fig. 23 on page 35

Images for this section:

Fig. 6: 46-years-old female studied by sinonasal polyposis. Incidentally an extra-axial lesion in the preptone cistern with retroclival location and lobed morphology was identified in MRI, with the following features of image: A and B: Axial and sagittal CT images showed slight remodeling and posterior cortical irregularity of clivus contour (red arrows). C, D, E: Axial, coronal and sagittal T2-weighted images, demonstrated a lesion with isointense and hyperintense areas (green arrows) compatible with Physaliforma echordosis. F: This feature in the axial post contrast T1-weighted sequence showed absence of enhancement. Note basilar artery (orange arrow), which has a posterior location regarding to lesion referred.
Fig. 7: A, B: Axial unenhanced CT images of the skull base demonstrate a soft-tissue mass in the sellar area with lytic bone destruction that affects to sphenoid body, clivus, left petrous apex and dorsum sellae. Typical trabecular entrapment is seen (green arrow). C: Axial enhanced CT image focused on the sellar area shows the mass with minimal enhancement that extends into the prepontine cistern (yellow arrow) and into the left cavernous sinus with encasement of left internal carotid artery (blue arrow).
Fig. 8: 38 years-old male with occipito-cervical headache for 7 months. A: Axial CT scan of the skull base demonstrates bone sequestra at a lytic clival mass (green arrow). B: Sagittal reformatted CT scan reveals the lesion with an extension into the prepontine cistern with dystrophic calcification (yellow arrow).
**Fig. 9:** Coronal T1-weighted MR image shows a large, isointense mass in the left side of the clivus with upper extension to dorsum sellae. Laterally, the lesion grows into the left cavernous sinus and displaces the temporal lobe without invasion of the dural layer (red arrow) and the middle cranial fossa. There is lower extension into left Meckel cave and foramen ovale (blue arrow).
Fig. 10: 53 year old woman with a history of resection of sellar chordoma, has decreased right visual acuity and VI cranial nerve palsy. Coronal T1-weighted MR image demonstrates a right parasellar mass predominantly hypointense that has nodular hyperintense foci, a finding that may correspond to intratumoral hemorrhage or highly proteinaceous material.
Fig. 11: Axial T2-weighted MR image shows an intracranial chordoma with high signal intensity.
**Fig. 12:** Patient with recurrent chordoma in C2 vertebral body with upper extension to the distal clivus. Sagittal T2-weighted fat-suppressed MR image demonstrates a mass with low-signal-intensity septations that separate high-signal-intensity lobules ("dirty cauliflower"). There are hypointense intratumoral areas that are secondary to chronic hemorrhage, bone fragments, or highly proteinaceous mucoid material (yellow arrow). The mass extends anteriorly with displacement the posterior wall of the oropharynx. It has a posterior extension into spinal canal with displacement the spinal cord that shows a normal signal intensity.

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**Fig. 13:** A: Axial unenhanced T1-weighted MR image shows an iso-/hypointense mass in the right side of the clivus that extends into parasellar area and petrous apex. B and C: Axial and coronal contrast-enhanced T1-weighted MR images show the mass with marked enhancement.

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Fig. 14: Axial unenhanced T1-weighted MR image shows a hypointense mass in sphenoid bone and clivus that extends anteriorly into the sellar/suprasellar area and laterally into both cavernous sinuses, more the left. B: Coronal T2-weighted MR image shows the mass with a heterogeneous signal intensity, predominantly hyperintense, that displaces and compresses upward pituitary gland and optic chiasm. C: On axial contrast-enhanced T1-weighted MR image the mass exhibits little and variable enhancement (necrosis or mucinous material in the tumor).
Fig. 15: Mass focused at the central skull base, in the sella turcica and dorsum sellae. A: Sagittal T1-weighted MR image shows the tumor with a heterogeneous signal intensity, with hypointense and hyperintense areas. The mass has a posterior extension into prepontine cistern with altered contour of the anterior wall of the pons (green arrows) and an anterior extension into sphenoid sinus (blue arrow). B: Coronal T2-weighted MR image demonstrates a multiseptate, predominantly hyperintense mass involving laterally to both cavernous sinuses, especially the right side. Note a vascular tubular structure with complete encasement by mass in relation to a persistent trigeminal artery (red arrow). C: Sagittal contrast-enhanced T1-weighted MR image reveals a heterogeneous and perifocal enhancement with multiple septa and internal loculations. D: lateral view of the right internal carotid arteriogram demonstrates a persistent trigeminal artery extending from the right internal carotid artery to the basilar artery (yellow arrow). E: Coil embolization of persistent trigeminal artery.
Fig. 16: Large mass in the central skull base with involvement of clivus, left petrous apex, yuxtaselar region, cerebellopontine angle and extension into left internal auditory canal. A. Coronal T2 weighted sequence of MRI shows lesion with marked hyperintensity of signal. B. CT demonstrates extensive lytic mass in the clivus extending to both sides of the midline and involvement of the left petroclival suture and the apex of the left temporal petrous (green circle). C, D, E. Postcontrast T1-weighted sequence of MRI in axial, sagittal and coronal planes shows intense and heterogeneous enhancement of lesion.
Fig. 17: Hemangioma located in left parasellar region. In the study of CT, lesion is slightly hyperdense compared to brain parenchyma. Contrast-enhanced CT shows marked and homogeneous enhancement. In MRI sequences the lesion is hypointense on T1 characteristically hyperintense on T2-weighted sequences. A slowly progressive contrast enhancement is evidenced in dynamic study of MRI, a hallmark of hemangioma.
**Fig. 18:** Central skull base metastasis. Axial, coronal and sagittal CT images show a mass centered in clivus with extensive cortical bone destruction associated (red arrows and green circle) and small intralesional calcifications/areas of microhaemorrhage (yellow arrow). Axial, coronal and sagittal FIESTA images demonstrate a hyperintense mass in clivus accompanied by prevertebral soft tissue mass (blue arrows). On axial postcontrast T1-weighted MR image an homogeneous and intense enhancement is observed.
Fig. 19: Myeloma in clivus, MRI findings. Mass affecting the clivus with suprasellar extension. It is isointense in T1-weighted and T2 weighted sequences, unlike chordoma. It extends laterally into the right cavernous sinus (red arrow), anteriorly to sphenoid sinus and posteriorly to the dural sheet. In the dynamic study after injection of gadolinium, the lesion presents early enhancement and washing compared with the pituitary gland, which is displaced superiorly (yellow arrow).
Fig. 20: Central skull base meningioma. Axial CT shows an hiperdense mass infiltrating clivus with right extension into the cavernous sinus and associated bone sclerosis in the clivus. Coronal T2 and axial T1 weighted images through the central skull base region demonstrate a homogeneous isointense mass (red arrow). Axial and sagittal postcontrast T1-weighted images demonstrate homogeneous enhancement of the mass (yellow arrow).
Fig. 21: Macroadenoma, MRI findings. An expansive lesion that occupies sella turcica is observed, extending to sphenoid sinus and clivus. T1 and T2-weighted images demonstrate isointensity of the lesion and enhancing in postcontrast images.
**Fig. 22:** 9-years-old boy with fibrous dysplasia. Axial, sagittal and coronal images of CT and MRI, including a CT volume rendering. Extensive dense bone lesion (yellow star) insufflates the cortical, occupying the anterior skull base with extension to frontal sinus, ethmoid cells and middle and inferior right turbinates, affecting the clivus, parasellar region and both cavernous sinuses. The lesion is iso-hypointense on T1-weighted images (red arrows) and hypointense on T2-weighted image (green arrow).

**Fig. 23:** 48-years-old female HIV positive presented with fever and headache. MR images demonstrate a lesion centered in chiasmatic hypothalamus region, hypointense on T1, heterogeneous on T2 with hyperintense areas, which shows peripheral ring enhanced after administration of gadolinium. Postoperative biopsy determined the diagnosis of craniopharyngioma.
Conclusion

Knowledge of development, function and anatomic path of notochord during embryonic stage allows assessment of entities related to the notochord which are described in postnatal life. Spectrum of these entities include anatomic variants, benign tumors and malignant neoplasms. The locations and origins of these disorders are intimately linked to the functions of the notochord during embryonic period and specifically with their anatomical path through the skull base.

Ecchordosis physalifora is considered a benign tumor of notochordal cells, rare, which is easily recognizable by MRI, frequently as an incidental image. It arises in the same location that intradural extraosseus chordomas. Their similarities regarding to radiological and histological features complicate their differential diagnosis, although the clinical course, immunohistochemistry and contrast-enhanced MR images provide key data in their distinction.

Chordomas are malignant neoplasm derivated from notochorda. Study of notochordal growth facilitates the comprehension about possible locations acquired by chordomas in the craniocervical junction.

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