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SOCIETY FOR NEUROSCIENCE IN ANESTHESIOLOGY AND CRITICAL CARE

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Craniovertebral Junction (CVJ) Anomalies

Neuroanesthesia Quiz #85

Start

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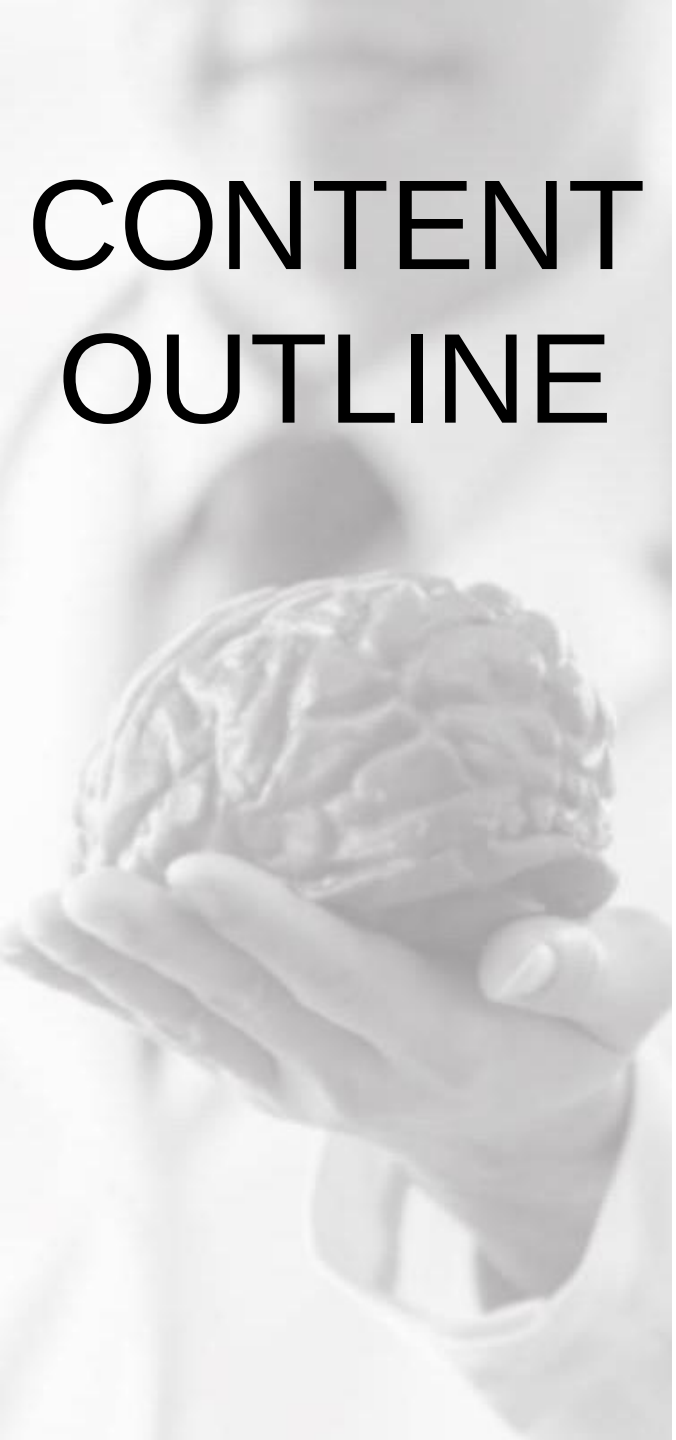
CONTENT OUTLINE



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QUESTION 1

Please click on any of the following links to proceed to that question/topic.

Which of the following statement is ***FALSE*** about Craniovertebral junction (CVJ) anomalies?

A: Congenital CVJ anomalies result due to defect in the development of occipital sclerotomes and adjacent cervical sclerotomes

B: The bony components of CVJ include occiput, foramen magnum, and the first cervical vertebra

C: The neural components of CVJ comprise of medulla, cervicomedullary junction, lower cranial nerves (IX, X,XI) and C1-C2 nerve roots

D: Major arteries traversing the CVJ include bilateral vertebral arteries, posterior inferior cerebellar arteries and meningeal branches of vertebral arteries

Content Outline

Q2, Q3, Q4, Q5

Sorry! Incorrect.

EXPLANATION

A. Congenital CVJ anomalies result due to defect in the development of occipital sclerotomes and adjacent cervical sclerotomes

This statement is true!!

In the early weeks of intrauterine life, the mesodermal cells form notochordal process, which invaginates between endoderm and ectoderm to form notochord. Somites are formed from the mesoderm on either side of the notochord. Sclerotomes are venteromedial portions of these somites and form the vertebral bodies.

Haldar R, Srivastava AK, Dikshit P, Jangra K. Anesthesia for Craniovertebral junction anomalies in pediatric patients. In. Fundamentals of pediatric neuroanesthesia. Rath GP (Ed). Springer Nature Singapore (2021). Pp 255-272

Offiah CE, Day E. The craniocervical junction: embryology, anatomy, biomechanics and imaging in blunt trauma. Insights Imaging. 2017 Feb;8(1):29-47.

Return to Question

Great Job!! Correct.

EXPLANATION

B. The bony components of CVJ include occiput, foramen magnum, and the first cervical vertebra

This statement is false!!

The bony components of the CVJ include the occiput, foramen magnum, the first two cervical vertebrae. The atlanto-occipital and atlantoaxial joints are all synovial joints.

Haldar R, Srivastava AK, Dikshit P, Jangra K. Anesthesia for Craniovertebral junction anomalies in pediatric patients. In. Fundamentals ofpediatric neuroanesthesia. Rath GP (Ed). Springer Nature Singapore (2021). Pp 255-272

Return to Question

Sorry! Incorrect.

EXPLANATION

C. The neural components of CVJ include medulla, cervicomedullary junction, lower cranial nerves (IX, X,XI) and C1-C2 nerve roots

This statement is true!!

The CVJ anomalies which could be of bony or soft-tissue involvement, lead to compression of these neural structures and result in the symptoms like pain, weakness, respiratory difficulty, vertigo etc.

Haldar R, Srivastava AK, Dikshit P, Jangra K. Anesthesia for Craniovertebral junction anomalies in pediatric patients. In. Fundamentals of pediatric neuroanesthesia. Rath GP (Ed). Springer Nature Singapore (2021). Pp 255-272

Return to Question

Sorry! Incorrect.

EXPLANATION

Next Question

D. Major arteries traversing the CVJ include bilateral vertebral arteries, posterior inferior cerebellar arteries and meningeal branches of vertebral arteries

This statement is true!!

In addition to the above, bilateral internal carotid arteries also remain anterior to and within 1-2 cm of the anterior end of the atlanto-occipital articulation.

Haldar R, Srivastava AK, Dikshit P, Jangra K. Anesthesia for Craniovertebral junction anomalies in pediatric patients. In. Fundamentals of pediatric neuroanesthesia. Rath GP (Ed). Springer Nature Singapore (2021). Pp 255-272

Return to Question

QUESTION 2

Craniovertebral junction (CVJ) anomalies could be congenital or acquired. Which of the following is ***TRUE*** regarding specific anomalies?

Please click on any of the following links to proceed to that question/topic.

A: In basilar invagination, the odontoid prolapses into a narrow foramen magnum and causes dorsal compression of cervicomedullary junction

B: Bony destruction is never seen in CVJ tuberculosis

C: Grisel syndrome involves subluxation of the atlanto-axial joint due to lax ligaments following infections of the head and neck

D: Chiari I malformation involves descent of the medulla, fourth ventricle and caudal vermis

Content Outline

Q1, Q3, Q4, Q5

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Sorry! Incorrect.

EXPLANATION

A. In basilar invagination, odontoid prolapses into a narrow foramen magnum and causes dorsal compression of cervicomedullary junction

This statement is false !!

In Basilar invagination, odontoid process prolapses into foramen magnum and causes ventral compression. It is often associated with foramen magnum stenosis. It is a congenital condition having syndromic association with Klippel-Feil syndrome, osteogenesis imperfecta, Down's syndrome etc.

Haldar R, Srivastava AK, Dikshit P, Jangra K. Anesthesia for Craniovertebral junction anomalies in pediatric patients. In. Fundamentals of pediatric neuroanesthesia. Rath GP (Ed). Springer Nature Singapore (2021). Pp 255-27.

Return to Question

Sorry! Incorrect.

EXPLANATION

B. Bony destruction is never seen in CVJ tuberculosis

This statement is false!!

There are various classification systems of CVJ tuberculosis based on clinic-radiological findings eg Lifeso (1987), Khandelwal (1991), Behari (2003), AIIMS system (2008), Goel (2016). There are subtle differences amongst these systems and generally, Stage II and Stage III CVJ tuberculosis involves minimal to marked bony destruction, respectively. (Chaudhary K, Pennington Z, Rathod AK, et al. Pathogenesis and Staging of Craniovertebral Tuberculosis: Radiographic Evaluation, Classification, and Natural History. Global Spine Journal. 2022)

The disease begins with inflammation of intervertebral joints and then involves the adjacent vertebral body and intervertebral disc space. When the avascular disc dies, there is vertebral narrowing and vertebral collapse. (Stout J. Bone and joint tuberculosis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. (Accessed on Dec 18, 2022.)

Return to Question

Great Job!! Correct.

EXPLANATION

C. Grisel syndrome involves subluxation of atlanto-axial joint due to lax ligaments following infections in the head and neck

This statement is true !!

Grisel syndrome is a condition of uncertain etiology and is characterized by a non-traumatic rotary atlantoaxial subluxation associated with a head and neck infection. Children are most frequently affected.

Park SH, Park SH, Lee SH. Grisel syndrome: pathophysiological evidence from magnetic resonance imaging findings. Ann Rehabil Med. 2013 Oct;37(5):713-6.

Return to Question

Sorry! Incorrect.

Next Question

EXPLANATION

D. Chiari I malformation involves descent of medulla, fourth ventricle and caudal vermis

This statement is false!!

Chiari malformations were classified by Hans Chiari in1891, into four groups:

Type I: abnormally shaped cerebellar tonsils that are displaced below the level of the foramen magnum

Type II: also known as Arnold-Chiari malformation: downward displacement of the cerebellar vermis and tonsils, a brainstem malformation with beaked midbrain on neuroimaging, and a spinal myelomeningocele

Type III: a small posterior fossa with a high cervical or occipital encephalocele, usually with displacement of cerebellar structures into an encephalocele

Type IV: now an obsolete term and describes cerebellar hypoplasia unrelated to other Chiari malformations.

Chiari 0 malformation: anatomic aberration of the brainstem (posterior pontine tilt, downward displacement of the medulla, low-lying obex) but with normally placed cerebellar tonsils

Chiari 1.5 malformation: is a CM-II like malformation without spina bifida

Khoury C. Chiari Malformations. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. (Accessed on Dec 3, 2022.)

Return to Question

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QUESTION 3

A 16-year-old male presented with headaches, nystagmus, ataxia and sleep related breathing difficulty. On evaluation, he was diagnosed with **Chiari Type I malformation (CM-I) with syringomyelia**. In this regard, all of the following statements are true *EXCEPT*:

Please click on any of the following links to proceed to that question/topic.

A: Headache in this condition is characterized as 'cough headache' and usually occurs at occipital or suboccipital location

B: There is a direct correlation between how low the cerebellar tonsils are (below the foramen magnum) and the clinical severity

C: Breathing difficulty could be because of both, the central and obstructive sleep apnea

D: Surgical decompression is commonly preferred over conservative management

Content Outline

Q1, Q2, Q4, Q5

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Sorry! Incorrect.

EXPLANATION

A. Headache in this condition is characterized as 'cough headache' and usually occurs at occipital or suboccipital location

This statement is true !!

CM-I should be considered as a potential etiology in secondary cough headache. It is postulated that Valsalva maneuvers leads to exacerbation of the pain by causing impaction of the cerebellar tonsils at the foramen magnum. As defined by the International Classification of Headache Disorders (ICHD-3), headache attributed to CM-I is precipitated by cough or other Valsalva maneuver, is occipital or suboccipital in location and lasts less than five minutes.

Khoury C. Chiari Malformations. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. (Accessed on Dec 3, 2022.)

Return to Question

Great Job!! Correct.

EXPLANATION

B. There is a direct correlation between how low the cerebellar tonsils are (below the foramen magnum) and the clinical severity

This statement is false !!

CM-I is characterized by cerebellar tonsils that are abnormally shaped and downwardly displaced below the level of the foramen magnum. The normal cerebellar tonsils may lie up to 3 mm below the foramen magnum in adults. In general, tonsils lying 5 mm or more below the foramen magnum on neuroimaging are considered to be consistent with a Chiari malformation, though there is no direct correlation between how low the tonsils are lying and clinical severity. With infants, tonsils as low as 6 mm below the foramen magnum can still be normal.

Khoury C. Chiari Malformations. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. (Accessed on Dec 3, 2022.)

Return to Question

Sorry! Incorrect.

EXPLANATION

C. Breathing difficulty could be because of both, the central and obstructive sleep apnea

This statement is true!!

Sleep related breathing difficulty in CM-I is typically defined as an apnea-hypopnea index (AHI) of > 5/h in adults and >1/h in children and it could be central, obstructive or mixed (both central and obstructive) sleep apnea.

Sleep-Related Breathing Disorders and the Chiari 1 Malformation. Leu RM. Chest. 2015 Nov;148(5):1346-52.

Return to Question

Sorry! Incorrect.

EXPLANATION

Next Question

D. Surgical decompression is commonly preferred over conservative management

This statement is true!!

Decompressive surgery is indicated for patients with CM-I who are clearly symptomatic with lower cranial nerve palsies, syringomyelia, myelopathy, cerebellar symptoms, or occipital cough headache

Khoury C. Chiari Malformations. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. (Accessed on Dec 3, 2022.)

Return to Question

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QUESTION 4

Please click on any of the following links to proceed to that question/topic.

A 74-year-old female with rheumatoid arthritis (RA) presented with atlantoaxial subluxation (AAS). Regarding rheumatoid craniovertebral junction anomalies, which of the following is *FALSE*:

A: The risk of AAS is greater in patients with severe RA and seropositive status than milder disease and seronegative status

B: Acquired or secondary basilar invagination is also known as basilar impression

C: In patients with severe RA, AAS is more common than involvement of sub-axial cervical vertebrae

D: Direct laryngoscopy is better tolerated with posterior or vertical subtype of AAS than anterior subtype

Content Outline

Q1, Q2, Q3, Q5

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Sorry! Incorrect.

EXPLANATION

A. The risk of AAS is greater in patients with severe RA and seropositive status than milder disease and seronegative status

This statement is true!!

The risk of AAS increases with time and the clinical symptoms of myelopathy (muscle weakness, altered sensation) can mimic other co-existing rheumatoid complications such as peripheral neuropathy, joint pain or disuse muscle atrophy.

As the disease progresses, synovial pannus (metabolically active tissue consisting of inflammatory and synovial cells) expands and further interfaces with cartilage and bone leading to joint destruction and erosion.

Samanta R, Shoukrey K, Griffiths R. Rheumatoid arthritis and anaesthesia. Anaesthesia. 2011;66:1146–1159

Return to Question

Sorry! Incorrect.

EXPLANATION

B. Acquired or secondary basilar invagination is also known as basilar impression

This statement is true!!

Basilar impression also referred to as atlantoaxial impaction or vertical cranial settling results from softening of the bone at the skull base secondary to diseases like rheumatoid arthritis, Paget's disease, osteomalacia, hyperparathyroidism, osteogenesis imperfecta, rickets, skull base infection etc

Donnally III CJ, Munakomi S, Varacallo M. Basilar Invagination. [Updated 2022 Nov 19]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK448153/

Return to Question

Sorry! Incorrect.

EXPLANATION

C. In patients with severe RA, AAS is more common than involvement of sub-axial cervical vertebrae

This statement is true!!

In RA, CVJ is more commonly involved (occipital condyles and atlanto-axis) than subaxial cervical spine. If there is disease at multiple levels, plain radiography classically demonstrates a ‘staircase’ appearance.

Samanta R, Shoukrey K, Griffiths R. Rheumatoid arthritis and anaesthesia. Anaesthesia. 2011;66:1146–1159

Return to Question

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Great Job!! Correct.

Next Question

EXPLANATION

D. Direct laryngoscopy is better tolerated with posterior or vertical subtype of AAS than anterior subtype

This statement is false!!

The most common subtype (**anterior AAS**) is worsened by C1/C2 flexion and therefore direct laryngoscopy should be tolerated. Here, C1 and the head tend to move as a unit, so that subluxation is worsened by the head moving anteriorly whilst the upper cervical spine is left behind, e.g., putting a pillow behind the head. A useful technique is to keep the upper cervical spine supported whilst the head is not moved anteriorly, e.g., using a doughnut head ring with a large enough hole to accommodate the occiput.

Posterior or vertical AAS poses the risk of spinal cord compression during C1/C2 extension, the movement of which occurs during direct laryngoscopy, which should therefore be avoided.

Anterior AAS results from laxity of transverse ligament induced by proliferative C1-C2 synovial tissue.

Posterior AAS occurs when odontoid process is fractured (from axis)/destroyed or when C1 ring is fractured

Samanta R, Shoukrey K, Griffiths R. Rheumatoid arthritis and anaesthesia. Anaesthesia. 2011;66:1146–1159

Return to Question

QUESTION 5

A 66-year-old female with history of breast cancer was diagnosed with metastatic disease of the atlantoaxial spine. Pertaining to metastatic disease of craniovertebral junction (CVJ), which of the following is *FALSE*:

Please click on any of the following links to proceed to that question/topic.

A: Rotational pain in the cervical spine is the most common symptom

B: Majority of metastases to the spine occur in the CVJ

C: Myelopathy is an uncommon presenting symptom

D: The gold standard imaging modality is magnetic resonance imaging (MRI)

Content Outline

Q1, Q2, Q3, Q4

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Sorry! Incorrect.

EXPLANATION

A. Rotational pain in the cervical spine is the most common symptom

This statement is true!!

The most common presenting complaint in this cohort of patients is cervical spine pain (in the form of mechanical pain and occipital neuralgia). Rotational pain is present in 90% of patients with CVJ involvement.

O'Sullivan MD, Lyons F, Morris S, Synnott K, Munigangaiah S, Devitt A. Metastasis Affecting Craniocervical Junction: Current Concepts and an Update on Surgical Management. Global Spine J. 2018 Dec;8(8):866-871.

Return to Question

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Great Job!! Correct.

EXPLANATION

B. Majority of the metastasis to the spine metastasis occur in the CVJ

This statement is false!!

The spine is the most common site for metastasis amongst the skeletal system. Of these the majority are found within the thoracic spine. Metastasis to the CVJ accounts for just 0.5% of all spine metastases.

O'Sullivan MD, Lyons F, Morris S, Synnott K, Munigangaiah S, Devitt A. Metastasis Affecting Craniocervical Junction: Current Concepts and an Update on Surgical Management. Global Spine J. 2018 Dec;8(8):866-871.

Return to Question

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Sorry! Incorrect.

EXPLANATION

C. Myelopathy is an uncommon presenting symptom

This statement is true!!

Myelopathy is an uncommon presenting complaint in this subset of patients and is observed in 0% to 22% of cases.

Poynton AR, Bilsky MH, Girardi FP, Boland PJ, Cammisa FP Jr. Metastatic disease of the cervical spine. In: McLain RF, Lewandrowski KU, Markman M, Bukowski RM, Macklis R, Benzel EC, eds. Cancer in the Spine: Comprehensive Care. Totowa, NJ: Humana Press; 2006:247-254

O'Sullivan MD, Lyons F, Morris S, Synnott K, Munigangaiah S, Devitt A. Metastasis Affecting Craniocervical Junction: Current Concepts and an Update on Surgical Management. Global Spine J. 2018 Dec;8(8):866-871.

Return to Question

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Sorry! Incorrect.

EXPLANATION

D. The gold standard imaging modality is magnetic resonance imaging (MRI)

This statement is true !!

The gold standard imaging modality in characterizing cervical spine tumors is magnetic resonance imaging (MRI), for both soft tissue and bone tumors.

Nuclear medicine can be employed to help distinguish between benign and metastatic tumors.

Moulding HD, Bilsky MH. Metastases to the craniovertebral junction. Neurosurgery. 2010;66(3 suppl):113-118.

O'Sullivan MD, Lyons F, Morris S, Synnott K, Munigangaiah S, Devitt A. Metastasis Affecting Craniocervical Junction: Current Concepts and an Update on Surgical Management. Global Spine J. 2018 Dec;8(8):866-871.

Return to Question

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THANK YOU !!

Return to Content Outline, Q1, Q2, Q3, Q4, Q5

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