Cerebral Ventricle: Congenital Lesions

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1. Holoproencephaly, the false answer is:

A. Alobar holoproencephaly results from merger of lateral ventricles.
B. SHH mutation is responsible for most of the abnormalities.
C. It is present in 1 in 15,000 live births.
D. Alcohol abuse has not been implicated as a cause.
E. It is present in 5% of patients with Smith-Lemli-Opitz syndrome.

Answer D

- Alcohol abuse is thought to selectively destroy midline cells at the early stage of development.

2. Coarctation, the false answer is:

A. Refers to apposition of two ventricular walls.
B. Focal coarctation can isolate a connatal cyst.
C. The incidence is less than 1%.
D. It results from inflammation or gliosis.
E. It occurs more commonly in the occipital horn of the lateral ventricle.

Answer D

- Coarctation is an error of development and does not result from inflammation nor gliosis.

3. Coarctation, the false answer is:

A. Often requires active treatment.
B. Resolves spontaneously by 2 months of age.
C. Connatal cysts occur anterior to the foramen of Monro.
D. Can be differentiated from subependymal cysts.
E. Are benign lesions.

Answer A

- Ventricular coarctation often resolves spontaneously and does not require active treatment.

4. L1 Syndrome, the false answer is:

A. It is inherited in an autosomal dominant manner.
B. Bilateral absence of the pyramids on imaging is pathognomonic.
C. Aqueductal stenosis may be a feature.
D. Patients often present with adducted thumb.
E. Sons are affected while daughters are carriers.

Answer A

- L1 Syndrome is inherited in an X-linked manner.

5. X-linked hydrocephalus, radiographic features, the false answer is:

A. Symmetric hydrocephalus with posterior horn dilation.
B. Large massa intermedia.
C. Hypoplastic cerebellar vermis.
D. Large quadrigeminal plate.
E. Rippled ventricular wall seen before VP shunt insertion.

Answer E

- The rippled ventricular wall follows ventricular peritoneal shunt insertion and is thought to be pathognomonic for X-linked syndrome.

6. X-linked hydrocephalus: management, the false answer is:

A. Ventricular peritoneal shunt improves neurologic outcome.
B. There is no genetic therapy for L1CAM.
C. Medical termination of pregnancy may be a consideration.
D. Genetic testing is indicated in suspected patients.
E. Management is symptomatic.

Answer A

- Ventricular peritoneal shunt does not improve neurologic outcome but reduces head size and helps improve care of the baby by the caregiver.

7. Choroid plexus cysts, the false answer is:

A. Are usually bilateral and demonstrate signal characteristics similar to CSF.
B. Should be differentiated from choroidal metastasis in adults.
C. May be acquired or congenital.
D. Most are found incidentally.
E. Are usually benign.

Answer A

- Choroid plexus cysts are usually unilateral with signal different from CSF, an incidental finding that is often linked to trisomy 18.

8. Aqueductal stenosis, the false answer is:

A. Stenosis often occurs in the caudal one-third.
B. The adytum narrows to a dorsally based triangle shape.
C. The adytum is deformed into a round opening when the fourth ventricle is trapped.

D. A single ependymal layer lines the lumen of the aqueduct.
E. Results in raised ICP.

Answer B

- The adytum in the normal aqueduct has a characteristic dorsally based triangle that focally narrows to a funnel-like structure in stenosis.

9. Aqueductal stenosis, associated ventricular deformities, the false answer is:

A. Ventricular dilation.
B. Focal enlargement of the third ventricle.
C. Ventricular diverticula.
D. Spontaneous ventriculo-cisternostomies.
E. Periventricular cysts.

Answer E

- Subependymal cysts occurs as a result of separation of the ependymal from the subependymal layer.

10. Aqueductal stenosis, radiology features, the false answer is:

A. Enlarged inferior recess of third ventricle.
B. Abnormally thinned corpus callosum.
C. Fourth ventricle diverticulum.
D. Lateral ventricle diverticulum.
E. Ventriculomegaly in the setting of rhomboencephalosynapsis.

Answer C

- The diverticulum is found in the lateral ventricle.

11. Aqueductal stenosis, histopathologic classification, the false answer is:

A. Stenosis.
B. Not otherwise specified.
C. Forking.
D. Septum formation.
E. Gliosis.

Answer B

- NOS does not feature in the classification of aqueductal stenosis.

12. Dandy Walker Malformation: Definition, the false answer is:

A. It is a congenital malformation.
B. There is enlargement of the posterior fossa.
C. There is enlargement of the fourth ventricle.
D. The Dandy Walker variant has a normal size posterior fossa.
E. It can be an acquired disorder.

Answer E

- Dandy Walker Malformation and its variants are congenital malformations that the represent a continuum of developmental anomalies that are grouped together as Dandy-Walker complex.

13. Dandy-Walker Malformation: Epidemiology, the false answer is:

A. It is the most common cerebellar malformation.
B. DWM is more common in males.
C. It can be inherited as an autosomal recessive trait.
D. Gestational exposure to TORCH infections is a risk factor.
E. Incidence is 1 per 25.000 live birth.

Answer B

- DWM is more common in female by a ratio of 3:1.

14. Dandy-Walker Malformation: Pathology, the false answer is:

A. Hypoplastic fourth ventricle.
B. Hypoplastic vermis.
C. Some association with FOXC1 gene.
D. Atresia of the foramen of Lushka.
E. Enlarged fourth ventricle.

Answer A

- An enlarged fourth ventricle is the definition of DWM.

15. Dandy-Walker Malformation: epidemiology, the false answer is:

A. Prevalence is about 1 in every 30,000 births.
B. It can be diagnosed in adulthood.
C. Maternal use of warfarin is a risk factor.
D. It is three times more common in females.
E. There is no geographical distribution.

Answer B

- DWM is a congenital malformation that is present at birth, the ability to have a fetal diagnosis allows the determination of whether the condition is DWM or a simple hydrocephalus due to trapped fourth ventricle.

16. Dandy-Walker Malformation, Associations, the false answer is:

A. Hydrocephalus in 80–90% of patients.
B. Posterior fossa anomalies.
C. Encephalocele in 20–70% of cases.
D. PHACES syndrome.
E. Myelomeningocele.

Answer E

- Myelomeningocele is commonly associated with an Arnold Chiari type II malformation, not with DWS.

17. Dandy-Walker Syndrome: Evaluation, the false answer is:

A. Neuropsychological tests are not required.
B. Contrast MRI is the test of choice.
C. US is a useful initial investigation.
D. Vermian abnormalities are characteristic.
E. Physical examination may reveal neurocutaneous melanosis.

Answer A

- All patients should be assessed for their cognitive level of functioning to direct further care. 8–10% of patients with DWM have neurocutaneous melanosis.

18. Congenital causes of hydrocephalus, the false answer is:

A. Aqueductal stenosis.
B. Type 1 Chiari malformation.
C. Dandy-Walker cyst.
D. Platybasia.
E. Germinal matrix hemorrhage.

Answer E

- Germinal matrix hemorrhage occurs after delivery in premature babies.

19. In aqueductal stenosis, the false answer is:

A. The posterior fossa is small.
B. Hydrocephalus can be manifest in utero.
C. Bulging fontanelle is not a feature.
D. MacEwan's crackpot sign can be elicited.
E. Setting sun eyes is a feature.

Answer C

- Bulging fontanelle is a feature of hydrocephalus and is a prominent feature in aqueductal stenosis.

20. Arachnoid cyst, the false answer is:

A. Can cause hydrocephalus by obstruction of CSF in the third ventricle.
B. Simple cyst that does not secrete CSF.
C. Comprise 1% of intracranial masses.

D. Suprasellar cyst with hydrocephalus can present with macrocephaly.
E. Shunting of cyst is probably the best overall treatment.

Answer B

- Histologically, simple cysts are lined with cells capable of secreting CSF.

21. Hydranencephaly, the false answer is:

A. Is a neurulation defect.
B. Is a post neurulation defect.
C. Is rarely associated with facial dysmorphism.
D. Most commonly due to bilateral ICA infarcts.
E. May mimic maximal hydrocephalus.

Answer A

- Hydranencephaly is a post neurulation defect and thought to be due to bilateral ICA infarcts.

22. Hydranencephaly differs from hydrocephalus, the false answer is:

A. Hydrocephalus may respond to shunting.
B. EEG shows no cortical activity in maximal hydrocephalus.
C. Frontal lobe and frontal horns are not visible on CT in hydranencephaly.
D. Flow void in supraclinoid carotid on angiogram may be seen in hydranencephaly.
E. Transillumination is not a useful differentiating factor.

Answer B

- EEG is one of the best ways to differentiate the two conditions, it shows electrical activity in hydrocephalus but not in hydranencephaly.

23. Holoprosencephaly, the false answer is:

A. Is caused by failure of cleavage of telencephalon.
B. Associated with trisomy 13 in 80% of cases.
C. Most children do not survive beyond infancy.
D. The risk is increased in subsequent pregnancies.
E. Facio-cerebral dysplasia is uncommon.

Answer E

- Also called arhinencephaly due failure of cleavage of telencephalon vesicle, the degree of craniocerebral dysplasia mirrors the severity of the holoprosencephaly, it is a common feature.

24. Chiari 1. Investigations, the false answer is:

A. Craniocervical junction abnormalities can be seen on plain X-ray.
B. Non-contrast CT scan is a useful first investigation.
C. MRI of brain and C-Spine is the test of choice.
D. Cine MRI may demonstrate CSF blockage at the foramen magnum.
E. Myelography coupled with CT shows good reliability.

Answer B

- CT scan is poor at evaluating the craniocervical junction due to bony artifact, also in young children there is a need to limit radiation exposure.

25. Aqueductal stenosis, etiology, the false answer is:

A. Point mutation of the Xq28 locus (L1CAM gene).
B. Intrinsic pathology.
C. Unknown.
D. Use of warfarin.
E. Infection.

Answer D

- Warfarin has been implicated in the development of Dandy-Walker Malformation but not directly linked to aqueductal stenosis.

26. Dandy-Walker Malformation: Symptoms, the false answer is:

A. Seizures in 15%.
B. Cognitive impairment in 50–70%.
C. Decreased ICP because of increased posterior fossa size.
D. Impaired motor function.
E. Macrocephaly in 80%.

Answer C

- Raised ICP is frequent as hydrocephalus is associated with the condition in 70–90% of cases.

27. Chiari 1, management, the false answer is:

A. Early surgery is recommended.
B. Asymptomatic patients should be operated.
C. Cerebellar symptoms improve after surgery in 87% of patients.
D. Weakness symptoms do not respond well to surgery.
E. C1-C3 laminectomy is often required.

Answer B

- Asymptomatic patients can be followed up and surgery should be offered only when a patient develops symptoms attributable to the malformation.

28. Chiari 1, associations, the false answer is:

A. Syringomyelia in 3%.
B. Hydrocephalus in 9%.
C. Craniosynostosis.
D. Klippel-Feil syndrome.
E. Basilar invagination.

Answer A

- Syringomyelia is a prominent feature of Chiari 1 and is present in 30–70% of cases.

29. Chiari 1, epidemiology, the false answer is:

A. Females more than males.
B. Average age at presentation is fourth decade.
C. Symptoms last between 1 month and 20 years.
D. Spontaneous improvement usually occur.
E. Achondroplasia is an associated condition.

Answer D

- Even though natural history is still a matter of debate, patient may remain static for years, rarely do they improve spontaneously.

30. Chiari 1, Signs, the false answer is:

A. Upbeat nystagmus.
B. Foramen magnum syndrome in 22%.
C. Central cord syndrome in 65%.
D. Cerebellar signs in 11%.
E. Normal neurology in 10%.

Answer A

- Downbeat nystagmus is characteristic of Chiari 1, it indicates a structural lesion in the posterior fossa especially at the cervicomedullary junction.

31. Chiari 1, the false answer is:

A. It is also called primary cerebellar ectopia.
B. Disruption of CSF flow at the foramen magnum.
C. It is always a congenital lesion.
D. Displacement of the medulla is unusual.
E. Hydrocephalus is rarely present.

Answer C

- Chiari 1 can be congenital or acquired, any cause of raised ICP or lumbar decompression can precipitate a Chiari 1 malformation.

32. Dandy-Walker Malformation, treatment, the false answer is:

A. Shunting of posterior fossa.
B. Concomitant shunting of lateral ventricle.
C. ETV when aqueduct is patent.
D. ETV when aqueductal stenosis.
E. Observation for asymptomatic patients.

Answer D

- ETV is not recommended in the presence of aqueductal stenosis.

33. Schizencephaly, the false answer is:

A. Unlike porencephalic cyst, it does not communicate with the ventricle.
B. Typically, it is lined with cortical grey matter.
C. It always communicates with the ventricle.
D. Septum pellucidum is absent in 80–90% of cases.
E. Seizure is a common presenting symptom.

Answer A

- Both porencephalic and schizencephaly may communicate with the ventricle, schizencephaly always, porencephaly sometimes.