Classical Signs and Appearances in Pediatric Neuroradiology

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Learning objectives

1-To illustrate 28 classic signs and appearances in pediatric neuroradiology.

2-To discuss the causes for their appearances, and the differential diagnoses.
Background

To correctly identify pediatric neuroradiological cases, radiologists should be familiar with the imaging characteristics and understand pathophysiological origins of these cases. Pediatric neuroradiologic glossary has been gradually enriched with the explanations of the imaging findings commonly used in daily practice. Classical signs and appearances explaining these imaging findings assure radiologists about diagnosis. In this review, classical appearances and signs common to neuroradiology practice will be defined. Pathologies causing these appearances and signs and possible differential diagnoses will be discussed.
Findings and procedure details

Classical Signs and Appearances

1. **Ice-cream cone sign:**

It reflects normal appearance of incudo-malleolar joint formed by malleolar head and body of incus on the axial computed tomography (CT) section (Figure 1). Anatomical identification of this anatomic structure is important in terms of ossicular luxation especially in trauma cases. The space between the ice-cream cone and the scutum is called Prussak's space.

2. **CT reversal sign:** The reversal sign is associated with diffuse anoxic-ischemic brain damage and almost always observed in children (Figure 2). This sign is characterized by a relative reversal of the attenuation between supra- and infratentorial structures. Gray-white matter distinction is lost and decreased, and there is a diffuse decrease in density in cerebral gray and white matter. Thalami, brainstem, and cerebellum have a relatively increased density. It is closely related to child abuse when it especially accompanies intracranial bleeding. \[1, 2\]

3. **Mount Fuji Sign:** This sign is observed in bilateral subdural tension pneumocephalus. These air accumulations lead to compression in frontal lobes and take a form of Mount Fuji on axial CT sections (Figure 3). It is most commonly seen after surgical decompression of chronic subdural hematoma. However, it may also be observed following head trauma, otogenic infections, nitrous oxide anesthesia, and diving. \[3\]

4. **Lemon sign:** The lemon sign is useful in identification of spina bifida and is commonly associated with hydrocephalus and Chiari II malformation. Loss of normal convex contour of frontal bones in transverse fetal sonogram obtained at biparietal diameter (Figure 4). It has a high sensitivity and specificity in high-risk patients before 24th gestational week. However, it is not specific to spina bifida and may be detected in encephalocele, Dandy-Walker malformation, tanatophoric dysplasia, cystic hygroma, corpus callosum agenesis, hydronephrosis, and umbilical vein varices. \[4\]

5. **Pancake brain sign:** This sign defines the appearance of abnormal brain tissue in cases with alobar holoprosencephaly. Holoprosencephaly is an anomaly caused by a prosencephalic division defect and characterized by varying degrees of fusion of cerebellar hemispheres, diencephalon, basal ganglia, and thalami. Pancake brain sign is formed by fusion of cerebral hemispheres associated with the presence of typical monoventricle at the center (Figure 5). \[1\]

6. **Molar tooth sign:** Joubert syndrome is an autosomal recessive disorder characterized by abnormal eye movements, nystagmus, and difficulty in following mobile objects
with eyes, apnea-tachypnea episodes, and motor retardation.\cite{5, 6} Molar tooth sign represents the abnormal antero-posterior orientation of superior cerebellar peduncles in a way similar to stems of a molar tooth on axial CT or magnetic resonance (MR) images (Figure 6). It is mainly observed in patients with Joubert syndrome.

### 7. Figure Eight Sign
Lissencephaly is a disorder caused by a defective neuronal migration between 8-14th gestational weeks and characterized by lack of development of gyri and sulci. Lissencephaly is classified into two subgroups: complete (type 1-agyria) or partial (type 2-pachygyria). Type 1 lissencephaly is characterized by shallow sylvian fissures that are vertically oriented. In this type of lissencephaly, brain takes on an hour glass or figure-8 appearance due to compression at the middle part by sylvian fissures on axial imaging (Figure 7).\cite{7}

### 8. Face of the Giant Panda Sign
This sign was first described by Hitoshi et al. in Wilson's disease in 1991. Consists of high signal intensity in the tegmentum except for red nucleus, preservation of signal intensity at the lateral portion of the pars reticulata of the substantia nigra, and hypointensity of the superior colliculus (Figure 8). The real pathology responsible from this appearance is the paramagnetic effect of the accumulation of heavy metals such as iron and copper in affected sites.\cite{8, 9}

### 9. Radial Band Sign
Radial bands are linear or curvilinear areas with an abnormal signal intensity extending from periventricular region to subcortical region, that are best observed on T2-weighted (T2W) and especially FLAIR MR sections (Figure 9). It is believed that radial band sign is indicative of abnormal migration of dysplastic stem cells during the course of radial glial-neuronal unit in patients with tuberous sclerosis complex. Radial bands are seen hypo-/iso-intense on T1-weighted images and hyperintense on T2W and FLAIR images.\cite{10}

### 10. String Sign/Tigroid (Leopard Skin) Appearance
This sign is characterized by multiple dark-spots or stripes (spared perivascular white matter) of normal white matter intensity located scattered within bright demyelinated periventricular white matter on T2W images (Figure 10). Tigroid appearance of the white mater has been defined in some cases with Pelizaeus-Merzbacher disease and metachromatic leukodystrophy. However, it has been recently reported that it may be observed in cases with lissencephaly accompanied by cerebellar hypoplasia.\cite{11, 12}

### 11. Open Circle Sign
The open ring sign is a relatively specific sign for demyelination, helpful in distinguishing between ring enhancing lesions. It is observed in patients with multiple sclerosis. It is observed as a lesion showing contrast effect as a circle that incompletely encircles a demyelinated plaque. The lesion is a high-intensity one on T2W images and it may be difficult to distinguish from an abscess or astrocytoma in this form (Figure 11).

### 12. Light Bulb Sign
Diffusion-weighted (DW) MR imaging is the method that can delineate ischemic lesions in brain at the earliest stage. With the help of this method,
lesion can be demonstrated within after the onset of the event. Ischemic area shines like a light bulb at this stage (it appears darker on ADC images) (Figure 12). This area forms the core of the infarcted region. The brightness diminishes by 2-3 months. By this way, acute and chronic infracts can be distinguished or acute lesions can be defined in those with multiple lesions of varying age. The marked increase in DWI signal in areas of acute ischemia, relative to unaffected brain, is typically so striking that this finding has been referred to as the "light bulb sign" of acute stroke.

13. **Keyhole sign:** The posterior fossa dimensions are normal in Dandy-Walker variants. There is a mild vermian hypoplasia and thus vallecula becomes widened between cerebellar hemispheres under the vermis. Fourth ventricle and cisterna magna communicates with each other through this wide vallecula. This appearance on axial CT and MR images is called keyhole sign (Figure 13).

14. **Dawson finger:** It is detected on MR examination in multiple sclerosis. Demyelinating plaques are observed as focal signal areas on proton density and T2W MR images (Figure 14). These plaques are round or ovoid lesions limited particularly to periventricular region. The appearance of periventricularly located ovoid lesions in the extended form along the ventricle is called Dawson finger.

15. **Cortical vein sign:** This was first described in MRI and also reported later on US and CT. It is used to differentiate extra-axial located subarachnoid and subdural effusions from each other. On both CT and MRI, bridging veins extend from cortical surface to arachnoid (Figure 15). Appearance of bridging veins coursing in that manner in an extra-axial fluid is called as positive cortical vein sign and indicates that the fluid is subarachnoid located. The fluid is subdural located when these veins are invisible.

16. **Caput medusa sign:** The most common vascular malformation in brain is venous angiomas. They are most commonly observed in frontal lobe and posterior fossa. It has been suggested that they stem from a pause at a time during brain development when arterial system has completed its development but venous system has not fully developed yet. The caput medusa sign, also known as a *palm tree sign*, refers to developmental venous anomalies of the brain, where a number of veins drain centrally towards a single drain vein. (Figure 16). The appearance is reminiscent of Medusa, a gorgon of Greek mythology, who was encountered and defeated by Perseus. The sign is seen on both CT and MRI, when contrast is administered.

17. **Angel wing sign:** Chiari type II is the most common type of Chiari malformation. It is also known as Arnold-Chiari malformation. In 90% of cases there is also myelomeningocele, hydrocephalus, and corpus callosum agenesis. In these cases, prepontine migration of the cerebellum at the level of the middle cerebellar peduncle gives the brainstem an angel wing appearance on axial MR images (Figure 17).

18. **Worm bag sign:** Arteriovenous malformations are space-occupying lesions formed by conglomerated large vessels. There may sometimes be a very small amount of brain
tissue between the vessels in intracranial located arteriovenous malformations. There is no brain tissue at all in some cases. Thus, such an appearance of large vessels resembles clustered worms and is called as worm bag sign (Figure 18).

**19. Tectal beaking:** Chiari type II is the most common type of Chiari malformation. It is also known as Arnold-Chiari malformation. In 90% of cases there is also myelomeningocele, hydrocephalus, and corpus callosum agenesis. Variable degrees of fusion of the colliculi and tectum result in prominent beaking and inferior displacement of the tectal plate. In these cases, the appearance of the pointed tectum is called tectal beaking (Figure 19).

**20. Double cortex appearance:** Because of the early arrest of neuronal migration, a symmetric circumferential band of heterotopic gray matter is separated from the overlying cortex by a thin band white matter. On MRI, the brain appears to have a "double cortex" appearance (Figure 20). The condition is quite rare, found predominantly in females, and is occasionally familial with an X-linked dominant inheritance.

**21. Banana cerebellum sign:** The banana cerebellum sign is one of the many a notable fruit inspired signs, such as lemon sign. In neural tube defects, folding of cerebellum around posterior brain stem due to inferior traction of spinal cord causes cerebellum to take the form of a banana. It has been reported that it may be present in 57% of fetuses with neural tube defect. In fetal hydrocephalus, a cerebellar deformation is observed in conjunction with ventriculomegaly and deletion of cisterna magna. In these cases, cerebellum loses its normal central convexity and becomes compressed in parallel to occipital bone, resembling a banana (Figure 21).

**22. Viking helmet appearance:** The "viking helmet" appearance refers to the lateral ventricles in coronal projection in patients with dysgenesis of the corpus callosum. The cingulate gyrus is everted into narrowed and elongated frontal horns (Figure 22). Dysgenesis of the corpus callosum may be complete (agenesis) or partial and represents an in utero developmental anomaly.

**23. The Tram-Track Sign:** The tram-track sign is seen on skull radiographs as gyriform, curvilinear, parallel opacities that have the appearance of calcifications (Figure 23). A similar appearance can be seen at CT. Sturge-Weber syndrome is a rare neurocutaneous syndrome that includes a facial port-wine stain and associated leptomeningeal angiomatosis. Weber demonstrated the characteristic gyriform intracranial calcifications. Calcifications are often gyriform and curvilinear and are most common in the parietal and occipital lobes. Calcification can be more extensive, however, with frontal lobe and/or bilateral involvement. CT scans show calcifications in areas of atrophy.

**24. Diamond shaped fourth ventricle:** This appearance is seen in rhombencephalosynapsis. Rhombencephalosynapsis is a rare condition in which most cases are found in newborns and infants. Morphological findings are predominantly characterized by fusion of the cerebellar hemispheres and absence of the vermis with often associated supratentorial anomalies. The size of the fourth ventricle is variable and
in its axial plane it usually has a "keyhole or diamond shape" (Figure 24). This appearance is the result of dorsal and rostral convergence of the dentate nuclei, cerebellar peduncles and the inferior colliculi.

25. **Bat-wing 4th ventricle**: Bat wing 4th ventricle sign refers to the morphology of the fourth ventricle in the Joubert anomaly and related syndromes. The absence of a vermis with apposed cerebellar hemispheres give the fourth ventricle an appearance reminiscent of a bat with its wings outstretched. It is best demonstrated on axial imaging plane (Figure 25) and could be easily missed on sagittal and coronal images.

26. **Bat-wing appearance of sylvian fissures**: Glutaric aciduria type 1 (GA-1) is an autosomal recessive inborn error of lysine, hydroxylysine and tryptophan metabolism that results from a deficiency of glutaryl-CoA dehydrogenase. The most striking finding on brain imaging is the presence of very wide CSF spaces anterior to the temporal lobes and within the sylvian fissures (giving a "bat-wing" appearance). Widening of the sylvian fissures is a very characteristic finding in glutaric aciduria type I (Figure 26).

27. **Frog eye appearance**: Anencephaly is the most severe form of cranial neural tube defects (NTD) and is characterized by absence of cortical tissue (although brainstem and cerebellum may be variably present) as well as absence of the cranial vault. Morphological spectrum within anencephaly ranges from holocrania (severest form) to merocrania (mildest form). Anencephaly may be radiological detectable as early as 11 weeks. A "frog eye" appearance may be seen when seen in the coronal plane US or MR images due to absent cranial bone / brain and bulging orbits (Figure 27).

28. **Boxcar ventricle sign**: Huntington disease is an autosomal dominant neurodegenerative disease, especially common in young adults. It has a course characterized by cognitive, behavioral, and muscle coordination disorders. In these cases, there may be atrophy in basal ganglia, particularly in caudate nucleus. Consequently, widening may be seen in frontal horns of lateral ventricle. This particular appearance of frontal horns on multiplanar MR sections is called boxcar ventricle sign (Figure 28).
Fig. 1: High resolution, axial CT image demonstrating the ice-cream sign of the temporal bone. The sign represents the typical appearance of the malleoincudal joint.

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Fig. 2: CT reversal sign is observed due to diffuse cerebral anoxia in non-contrast CT examination.

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Fig. 3: Mount Fuji sign due to tension pneumocephalus is observed in axial plane CT sections (parenchymal and bone window)

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Fig. 4: Lemon sign is seen in frontal bones in a fetus with myeloschisis, as detected in an obstetrical US performed at 20th weeks of gestation.

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Fig. 5: Pancake brain appearance formed by monoventricle cavity and cerebral hemispheric fusion is seen in T1-weighted MR image in a case with alobar holoprosencephaly.

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Fig. 6: Molar tooth sign (star) at the level of pons and superior cerebellar peduncles coursing parallel to each other (arrows) are seen in T1-weighted MR section in a case with Joubert syndrome.

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Fig. 7: An appearance similar to figure eight due to lissencephaly in axial plane CT examination.

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Fig. 8: A giant panda face is observed in T2-weighted axial MR image in a case with Wilson syndrome.

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Fig. 9: Hyperintense radial bands extending linearly at the level of right cerebral hemisphere and a cortical tuber located at left parietal lobe in FLAIR MR image on axial plane in a case with tuberous sclerosis complex.

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**Fig. 10:** A tigroid appearance is observed at periventricular white matter in axial T2-weighted MR sections in a 2-year-old girl with metachromatic leukodystrophy.
Fig. 11: Post-contrast T1-weighted MR image showing incomplete ring enhancing lesion in the left parietal region.

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**Fig. 12:** This b=1000 s/mm² DWI showing the acute infarct as "light bulb" bright.

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**Fig. 13:** Axial non-contrast CT image showing typical "key-hole" appearance of cisterna magna communicating with dilated 4th ventricle.

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**Fig. 14:** Parasagittal FLAIR MRI demonstrating multiple sclerosis plaques extending up through corpus callosum.

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Fig. 15: Post-contrast axial CT image showing the cortical veins.

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Fig. 16: Contrast-enhanced T1-weighted axial MR image confirming converging tubular structures that represent a venous angioma in medial aspect of right cerebellar lobe.

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Fig. 17: Axial T2-weighted MR image showing an angel wing appearance in brainstem.

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**Fig. 18:** Sagittal T2-weighted MRI images showing a nidus of compact vessel with the typical appearance of "bag of black worms" in the left frontal region.

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Fig. 19: Sagittal T1-weighted MRI demonstrating small posterior fossa with low lying tentorial attachment posteriorly. The tectum is beaked and partial corpus callosum agenesis is present.

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**Fig. 20:** T2-weighted MR image demonstrating a "double cortex" appearance.
Fig. 21: Antenatal ultrasonogram showing a banana sign.

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Fig. 22: Coronal view of MRI head of the patient demonstrating the lateral ventricles forming a "Viking helmet" appearance due to the absence of corpus callosum.

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Fig. 23: Lateral skull radiograph in a patient with Sturge-Weber syndrome showing parallel cortical calcifications. Contrast-enhanced axial T1-weighted MRI showing gyriform contrast enhancement in the right cerebral hemisphere. There is brain atrophy on the right side. The cranial vault is asymmetric as secondary to brain atrophy.

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**Fig. 24:** Axial T2-weighted MRI at the level of the posterior fossa showing antero-posterior elongation of the fourth ventricle giving it a "diamond shaped" appearance.

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**Fig. 25:** Axial T1-weighted and T2-weighted images demonstrating widened sylvian fissures producing "bat-wings" appearance.

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Fig. 26: Fetal MR images demonstrating absent cranial bone / brain and bulging orbits. In addition, polyhydramnios is seen.

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**Fig. 27:** Axial and coronal T2-weighted MR images showing bilateral atrophy of caudate nuclei and compensatory dilatation of lateral ventricles, a finding known as "boxcar ventricle".

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Conclusion

The use of signs and appearances for interpretation of images in pediatric neuroradiology is quite useful. The majority of these signs and appearances are quite specific and, in many cases, pathognomonic.