Descriptive neuroradiology: beyond the hummingbird

Inna Page,1 Frank Gaillard 1,2

ABSTRACT
Radiology signs have long been described in ways that communicate the imagery around us to enhance our cognitive perception. Here, we describe the use and limitations of 10 such signs in neuroradiology, divided into three groups. The first are signs that are reliable for a specific diagnosis, such as the Medusa head sign indicating a developmental venous anomaly, or a racing car sign in agenesis of corpus callosum. The second group of signs helps us to diagnose rare conditions, such as the onion skin sign in Balo’s concentric sclerosis. The third group is of unreliable signs that may lead clinicians astray. For example, the absence of a swallow-tail sign in Parkinson’s disease or the presence of a hummingbird sign and Mickey Mouse sign in progressive supranuclear palsy. The appropriate use of these signs in clinical practice is essential.

INTRODUCTION
Over the last century, the art of radiology has become a key to diagnosing and differentiating between a myriad of neurologica condi- tions. Initially with plain radiographs, then angiography and CT scanning, and most recently MR imaging, radiologists have described the appearance of literally thousands of conditions. Many pathological appearances have been given memorable and sometimes whimsical names. This has led to a veritable Augean stable of radiological signs, such that otherwise dour radiologists can be heard muttering to their trainees about ‘hummingbirds’, ‘hot-cross buns’, ‘racing cars’ and ‘hockey sticks’. Essentially, radiological signs convey to the observer an appearance that demonstrates a certain distinctive pattern, which in turn helps the clinician to recognise characteristic abnormal findings or normal anatomical structures. Such signs may resemble flora and fauna, fruits and vegetables, landscapes and weather conditions, or even, unimaginatively, be named after the physician who discovered it.

Signs can be helpful because they facilitate clinician’s communication of findings to each other and can also increase a clinician’s confidence in making a specific diagnosis. Furthermore, in certain rare diagnoses with striking imaging appearances, radiological signs may be extremely helpful, as having been seen once they can be recalled to make an otherwise difficult diagnosis. This is known among radiologists as an ‘Aunt Minnie’: if you had a dear aunt named Minnie, then you would easily recognise her in a crowd of similar women, even though it would be difficult to articulate to someone who has never seen your aunt Minnie before exactly what features you are using to inform that recognition.

Although many of these signs seem to have genuine pedagogical and clinical utility, their importance is sometimes exaggerated, particularly by less experienced radiologists and clinicians, as they become thought of as pathognomonic and unduly relied upon for diagnoses. This, in turn, can lead the diagnostic process astray and result in delays or unnecessary tests or anxiety. Note that often these signs are little more than a familial lexicon handed down from one generation of radiologists to the next. Thus, there is scant literature on how they perform as clinical tests, with sensitivity, specificity and positive and negative predictive values generally not available. It is therefore essential to interpret them in the appropriate clinical context.

This article can cover only a small fraction of all described signs in neuroradiology, and so we have chosen 10 that illustrate the use, charm, limitations and dangers of these fickle creatures.

Medusa head sign and palm tree sign
Medusa head or caput Medusae sign of developmental venous anomalies is one of the most straightforward signs,
anecdotally rarely confused with other pathology, and genuinely useful in everyday practice. It is commonly identified on vascular imaging and comprises radially orientated medullary veins draining into a larger central vein, which in turn drains into the normal deep or superficial venous system (figure 1A). It takes its name from a Greek mythological Gorgon (figure 1B) out of whose scalp arose living snakes rather than hair. Unlike the dire consequence of looking at Medusa—becoming literally petrified—developmental venous anomalies are benign and almost always incidental and asymptomatic, although occasionally associated with cavernomas. If imaged longitudinally along their draining vein (figure 1C), their appearance is more reminiscent of a palm tree (figure 1D), in which case the associated cavernomas may resemble coconuts; some feel that this is pushing the analogy somewhat.

Racing car sign and moose head sign

Racing car sign is another straightforward aide mémoire that merely describes the appearance of the lateral ventricles on axial imaging in people with agenesis of the corpus callosum (figure 2A). The frontal horns are the front tyres and the trigones the back tyres. The body of the lateral ventricles is widely spaced by the white matter fibres that would normally cross the midline forming the corpus callosum, instead running inferiorly, medial to the ventricles, and known as the bundles of Probst. The importance of this sign is primarily historical as axial CT could easily overlook callosal agenesis. It now has far less utility with the availability of sagittal and coronal imaging routinely obtained as part of essentially all cross-sectional imaging (CT and MRI), as noting the absence of the corpus callosum is trivial. On coronal imaging, the appearance has instead been likened to a moose’s head (figure 2C, D).

**Figure 1** (A) Axial susceptibility weighted image (SWI) through the cerebellum demonstrates multiple veins draining centripetally towards a central draining vein (arrow) reminiscent of (B) head of Medusa, a Gorgon of Greek mythology. (C) Rotated axial SWI image along the draining vein has appearances strikingly similar to (D) a palm tree. Attributions: (A) Bruno Dimuzio, Radiopaedia.org rID: 41402. (B) Medusa by Michelangelo Merisi da Caravaggio or Caravaggio reproduced from commons.wikimedia.org/wiki/File:Medusa_by_Caravaggio.jpg (public domain), (C) Neil Lall, Radiopaedia.org rID: 46258, (D) Mark Basarab, Unsplash.com ID: eBoge2zCsr4.
Onion skin sign or bullseye sign

Some signs are useful in drawing attention to a particular feature of an abnormality allowing you to think of an otherwise uncommon condition. One such example is the onion skin sign or bullseye sign of Balo’s concentric sclerosis, a rare demyelinating disorder usually considered to be a variant of multiple sclerosis. The underlying pathological cause of this appearance is the presence of alternating layers of demyelinated and myelinated white matter resulting in concentric layers of high and low signal intensity on T1 and T2 weighted MR images (figure 3A). This appearance is considered to be pathognomonic for this condition.3 It is important, however, to note that the key to this appearance is the presence of multiple concentric rings, as a single ring may be seen in many other pathologies including cerebral abscesses and metastases.

Hockey stick sign

The hockey stick sign of variant Creutzfeldt-Jakob disease is another example of a sign helpful in identifying a fairly specific feature of a rare condition, thus markedly narrowing the differential diagnosis. This rare cause of dementia made the headlines in the 1990s due to its transmission from infected beef
Figure 4  Axial diffusion weighted image (A) shows abnormal diffusion restriction in the caudate nuclei (empty arrows), bilateral putamen (white arrows) and posterior medial thalami (open arrowhead). The latter resembles a hockey stick (B). Attributions: (A) Fateme Hosseinabadi, Radiopaedia.org rID: 73775 (B) Gerhard Crous, Unsplash.com ID: xi4aa21VA8.

due to an epidemic of bovine spongiform encephalopathy in cattle. This sign is appreciated best on axial diffusion weighted image images (figure 4A) but also faintly seen on fluid attenuation inversion recovery MR images. The combination of high signal intensity within the posterior and dorsomedial thalamic nuclei resembles a hockey stick (figure 4B). In the appropriate clinical context, the hockey stick sign is highly specific and sensitive for variant Creutzfeldt-Jakob disease, with one publication from 2000 reporting 78% sensitivity and 100% specificity.

FLAIR images in Wernicke’s encephalopathy may show a similar appearance, although the term hockey stick sign is not used in this context and white matter abnormality extends into the hypothalamus and mammillary bodies as well as periaqueductal grey matter.

Trident sign
Some signs are helpful as they prompt the reader to distinguish an unusual and important condition from far more common relatively benign findings. The trident sign is such an example. An ill-defined region of high T2 signal within the mid pons is common in people with chronic small vessel ischaemia. In contrast, the trident sign, a symmetric area of high signal intensity on T2 (figure 5A) and FLAIR axial images with three anterior projections is characteristic of osmotic demyelination (previously referred to as central pontine demyelination).

Given its shape, and a historical fad for naming signs after classical mythology, it is known as the trident sign referring to Poseidon’s weapon of war (figure 5B). One of the authors (FG) believes that this is actually far more reminiscent of the logo of the rebel alliance in Star Wars (figure 5D). Nevertheless, finding this sign, when supported by non-enhancing low signal intensity on T1 weighted images (figure 5C) and no mass effect, strongly suggests osmotic demyelination and can be used with confidence in the setting of a supportive clinical history.

Another potential shortcoming of catchy signs is that the same term is used in multiple contexts. For example, the trident sign of neurosarcoidosis describes the appearance of the spinal cord on the postcontrast T1 weighted axial images, enhancement of the central canal (centre prong) and the dorsolateral subpial enhancement (lateral prongs) also resembles a trident.7

Milky Way sign
Another example of this is the Milky Way sign. The literature describes it as one of the distinguishing clues to differentiate between progressive multifocal leukoencephalopathy (PML) (rare) and lesions of multiple sclerosis (common) in asymptomatic natalizumab-treated patients. Multiple star-like punctate foci of hyperintensity in the close vicinity of a larger lesion on T2 (figure 6) and FLAIR axial images are reminiscent of the Milky Way. In a 2016 publication, this sign was reported as 100% specific and 78% sensitive for PML.8 Of course, what we really need to know is its negative and positive predictive values. As usual, however, these cannot be gleaned from case series cohort and their utility in everyday practice is uncertain.

Noted that this punctuate pattern is not specific to natalizumab and can occur in PML from other causes. Nonetheless, it may serve as an additional imaging marker to increase our confidence in diagnosing PML in asymptomatic patients.9 10

Hot-cross bun sign
Another category of signs is those that are striking but seen only late in the course of a disease, at which time the diagnosis is often clinically obvious. In modern medicine practised in affluent societies where diagnoses are often made relatively early, these signs are therefore interesting but not very useful. The hot-cross bun sign is such a sign. It is best known for its association with cerebellar-type multiple system atrophy (MSA-C). Demyelination of the pontocerebellar fibres with preservation of the pontine tegmental tracts and corticospinal tracts results in a high T2 signal cross on the background of darker myelinated pons and will usually only be seen when there is substantial loss of pontine volume (figure 7).11 However, despite the previously documented high specificity of the hot-cross bun sign for MSA-C, a recent publication cast doubt on this, instead suggesting that it merely reflects pontocerebellar degeneration, MSA-C being the most common cause. In this study, although on a small sample, the authors reported only 6 out of the 11 cases meeting the clinical criteria for probable or possible MSA. The other causes included...
spinocerebellar ataxia, PML, bilateral middle cerebral peduncle infarction, variant Creutzfeldt-Jakob disease, fragile X tremor–ataxia syndrome and cerebrotendinous xanthomatosis. 

It is also important to note that an ill-defined cruciform appearance with reversed signal characteristics (dark cross or a light background) can also occur in chronic small vessel ischaemic change. Failure to remember this will result in gross overcalling of a rare condition.

Boxcar sign
Some signs are well known but no one seems to know what they actually mean, let alone have determined their utility with any degree of scientific rigour. Early presymptomatic Huntington’s disease has subtle or absent imaging signs. The imaging appearance later in the disease—when the caudate nuclei are volume-depleted, the widened and frontal horns squared off—may be referred to as the boxcar sign. The problem seems to be that we have collectively forgotten how this appearance is meant to suggest boxcars (figure 8B). Is it the appearance of the ventricles on coronal (figure 8A) or axial images? Is the brain meant to be the boxcar and...
the ventricles the windows? Or is it the outline of the ventricles each appearing rectangular and boxcar-like? Or something else? For that matter, we suspect many radiologists do not even know what boxcars are and the sign has rightly fallen out of favour due to the decline of railways and the ascendance of more reliable features, particularly the ratio of intercaudate distance to distance between the inner table of calvaria (CC:IT) and ratio of frontal horn width to intercaudate width (FH:CC) (Figure 8C). Note that the lentiform nuclei are at least as affected as the caudate nuclei; automatic morphometry (Figure 8D), which segments anatomical brain regions and calculates their volume, can elegantly demonstrate this. These findings are sensitive, but poorly specific.

Swallow tail sign
Some signs sound almost too good to be true, and sometimes they are. Idiopathic Parkinson’s disease has historically not had a reliable imaging biomarker. Then, the swallow tail sign was described as a normal

Figure 7  Axial T2 weighted image (A) demonstrates a cross-shaped hyperintensity in the atrophied pons giving an impression of a hot-cross bun (B). Attributions: (A) Frank Gaillard, Radiopaedia.org rID: 5465. (B) Jasmine Waheed, Unsplash.com ID: dRJGQAh6x8U.

Figure 8  Coronal T2 weighted image (A) demonstrates caudate nuclei (black outline arrow) and putaminal (white arrow) atrophy with secondary dilatation of the frontal horns of the lateral ventricles apparently resembling a boxcar (B). Axial T2 weighted image (C) demonstrates the intercaudate distance (red arrow), frontal horn width (yellow arrow) and inner table distance (purple arrow). Putaminal atrophy (white arrow). Axial morphometry (D) shows significant volume loss in basal ganglia, especially in putamen. Attributions: (A), (C) Mostafa El-Feky, Radiopaedia.org rID: 56374. (B) Claudio Mezzasalma, Unsplash.com ID: aANiWvNjXrQ. (D) Frank Gaillard, Radiopaedia.org rID: 55786.
appearance of the substantia nigra in a healthy population. The dorsolateral substantia nigra on SWI shows low signal intensity (figure 9A) and resembles a split tail (figure 9B) surrounding the high signal intensity of nigrosome-1. Patients with Parkinson’s disease and dementia with Lewy bodies were found to have lost the swallow tail sign; this was proposed as an indicator to facilitate the diagnosis of Parkinson’s disease. This sign is best appreciated on images using 7 T magnets and to a lesser degree 3 T magnets. However, there are some limitations in clinical applicability of this sign. One study showed 19% false-positive results in a healthy population. One of the main limitations in daily practice is the field strength. The higher the field strength, the higher the accuracy. However, the most commonly used scanners in daily practice are 1.5 T and 3 T, with 7 T scanners at present reserved mostly for research. The second limitation is that this sign is very sensitive to motion artefact, and non-motion degraded imaging can be challenging to achieve in many Parkinson’s patients. An additional cause for reduced accuracy is thought to be individual molecular variants of nigrosome-1. Due to variable appearance in the healthy population, need for higher field strength magnets and degradation by motion artefact, this sign should be used only cautiously in clinical practice. 

Hummingbird sign and Mickey Mouse sign
Lastly, some signs are useless, not only failing to help in making the correct diagnosis but going out of their way to muddy the diagnostic waters. The undisputed champion in this category is the hummingbird sign and the less well-known Mickey Mouse sign, supposedly imaging patterns of progressive supranuclear palsy. The hummingbird sign is seen on midsagittal images (figure 10A) due to selective midbrain tegmentum atrophy with relative preservation on the pons. Reduction in antero-posterior diameter of the midbrain and thinning of

![Figure 9](https://example.com/figure9.png)
the cerebral peduncles have a characteristic appearance on axial views (figure 10C) and are called the Mickey Mouse sign. Although the reported specificity of these signs is high, accurate recognition of these signs in early stages is challenging as the sensitivity is low.\textsuperscript{18 19} For example, a recent study found the hummingbird sign had a specificity of 99.5\% and a positive predictive value of 96.1\% but a low sensitivity at only 51.6\%.\textsuperscript{19} Therefore, whenever one thinks the midbrain resembles a hummingbird, it must be supported by quantified measurements, such as midbrain to pons ratio and Magnetic Resonance Parkinsonism Index in order to increase the diagnostic accuracy.\textsuperscript{20}

CONCLUSION
Radiological signs play an important role in radiology culture and aid the recognition of pathological findings. However, it is essential to apply them accurately in an appropriate clinical context.

FURTHER READING
1. Schott J M. ‘A neurological MRI menagerie’ covers some additional animal-inspired imaging signs. DOI: 10.1136/jnnp.2007.120261
2. Mulroy E. \textit{et al} ‘Animals in the Brain’ also focuses on animal-inspired signs but relates specifically to movement disorders.
REFERENCES