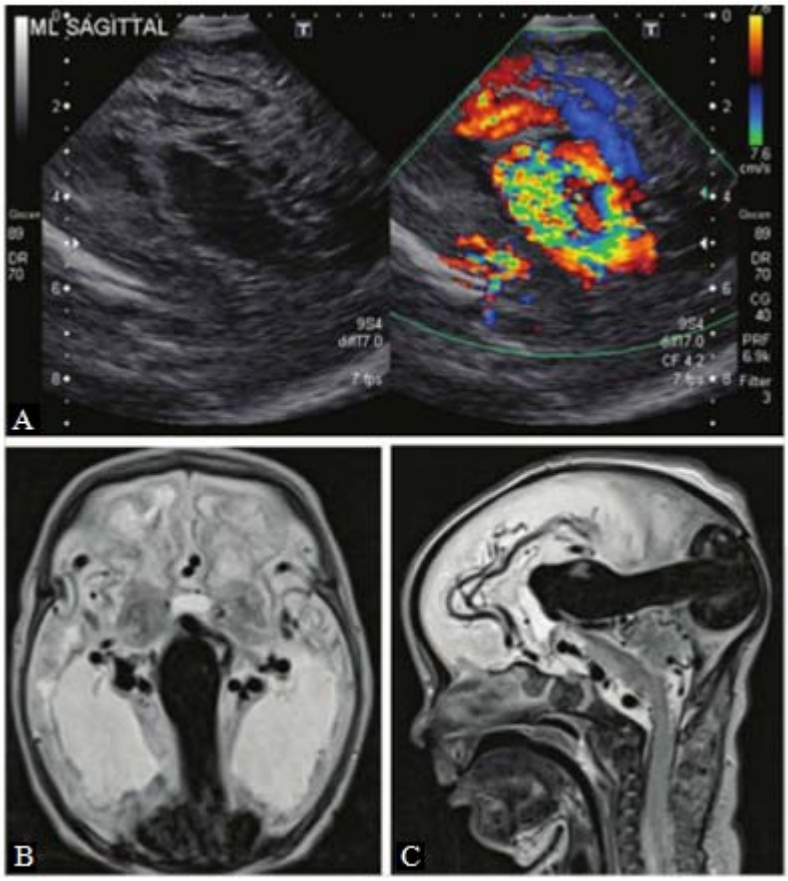
EANS/UEMS European examination in neurosurgery

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurosurgery, M.Gorky Donetsk National Medical University)

MISCELLANEOUS SCENARIO

Case 1

A 2-day-old boy with congestive heart failure.



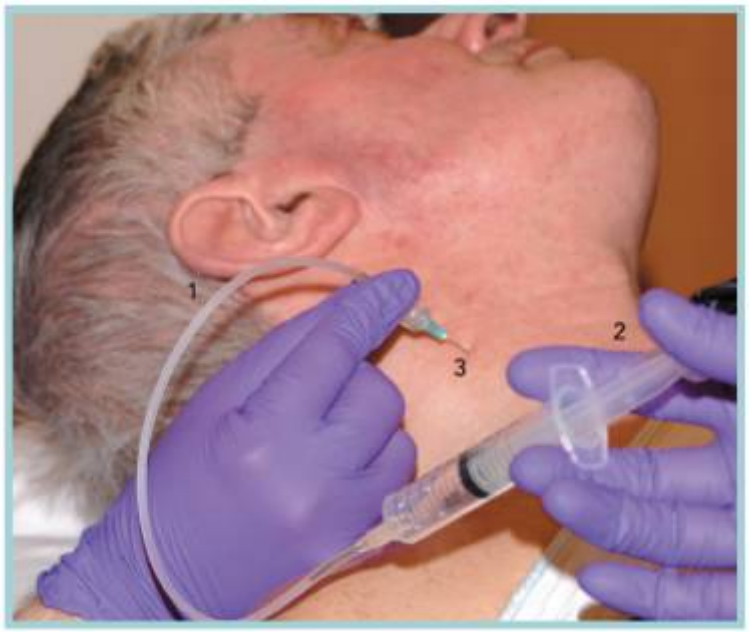
1. What are the imaging findings?
2. What is the most likely diagnosis?
3. What is classification for this pathology?
4. What is the best treatment for this boy?

Case 2

1. Draw a schematic diagram of the brachial plexus.
2. On your diagram show how the roots, trunk, divisions, cord and branches lie in relation to the bony landmarks of the neck and shoulder.

Case 3

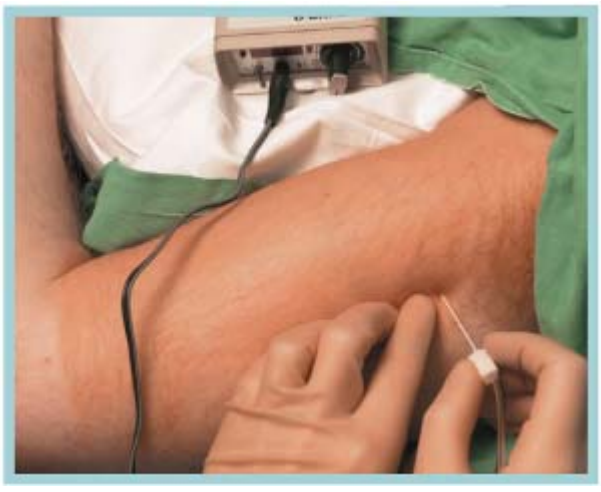
This patient is about to undergo carotid surgery under regional anesthesia using a deep cervical plexus block.



1. Is this block alone sufficient for this surgery?
2. What are the landmarks indicated by points 1, 2 and 3?
3. Which cervical dermatomes are blocked for this procedure to be successful?
4. Outline the anatomy of the procedure and how it is performed.
5. Is regional anesthesia preferable for this surgery?
6. What complications can occur using this regional block?

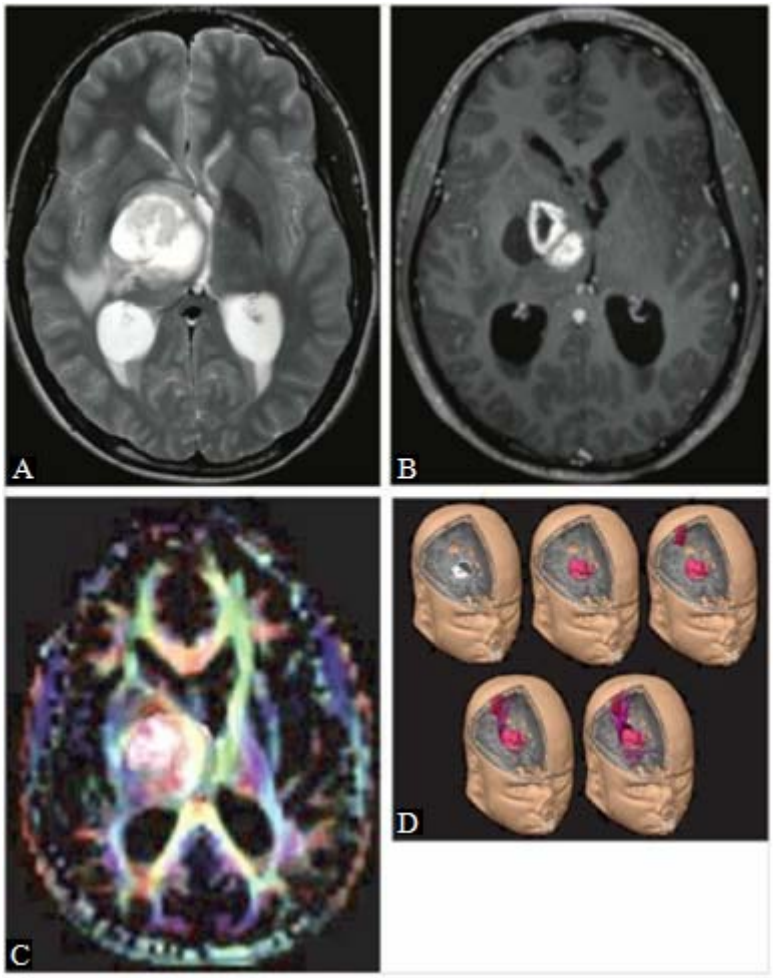
Case 4

1. What procedure is being shown?
2. How is accurate needle placement ensured?
3. Comment on the ‘transarterial’ technique.



Case 5

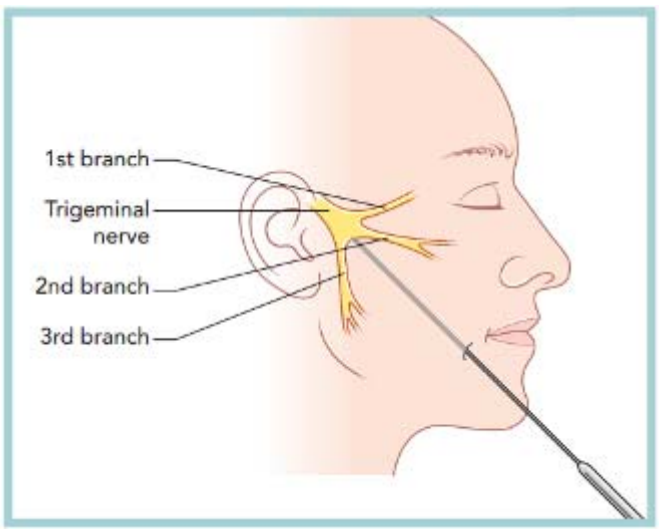
An adolescent boy with high-grade glioma referred for preoperative planning.



1. What are the imaging findings?
2. What is your diagnosis?

Case 6

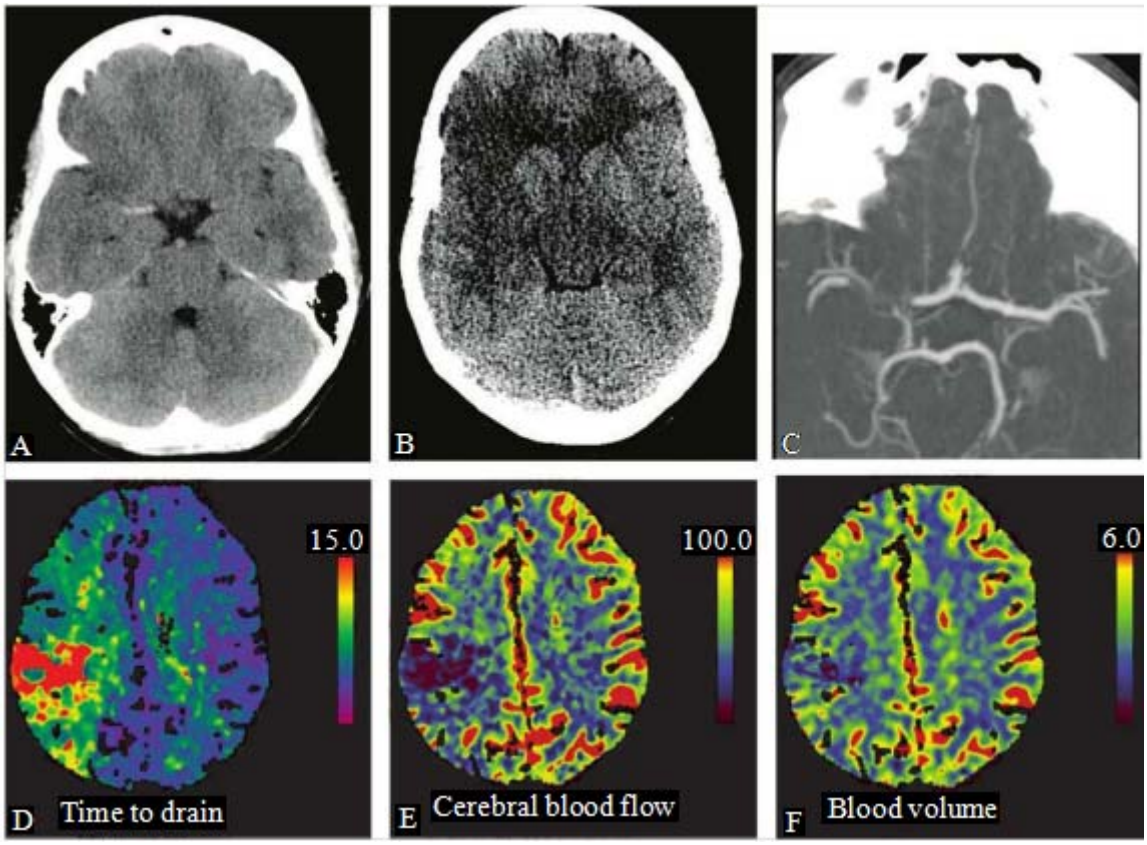
This patient has severe recurrent trigeminal neuralgia.



1. What technique is being carried out?
2. Why does it work?
3. Describe the anatomy of where the needle tip needs to lie and how this is safely achieved.

Case 7

Adult men with acute onset left-sided neurological deficits.

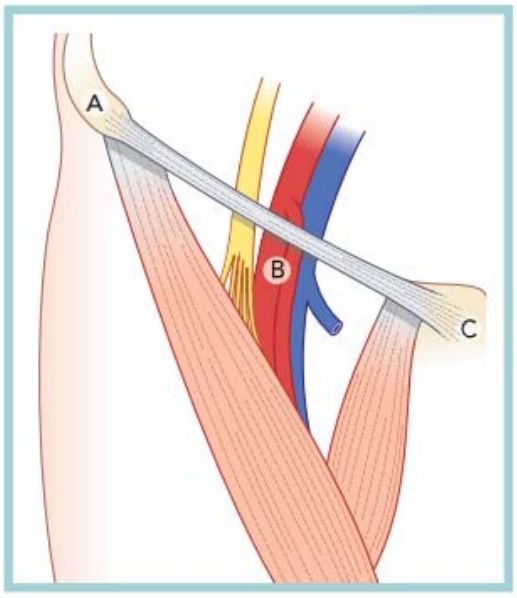


1. Describe the findings on the imaging study.
2. What is your diagnosis?
3. What is the best treatment for this patient?

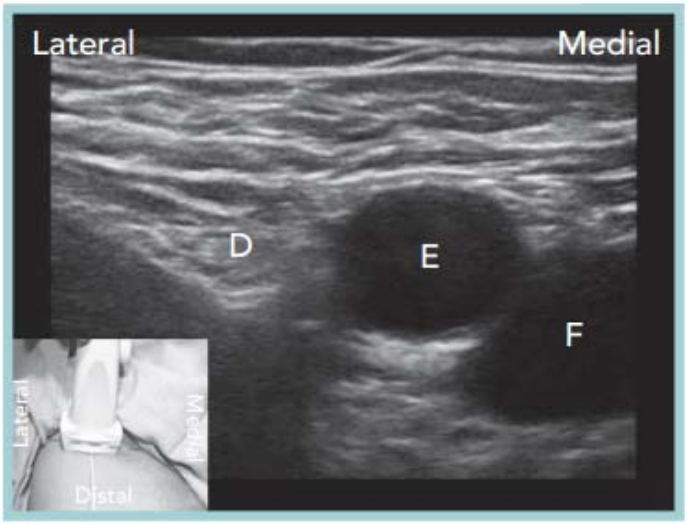
Case 8

A femoral nerve catheter for infusion of regional anesthesia is suggested for a 45-old-patient.

1. What structures, labelled A, B and C on Figure below, are used as landmarks to identify the approximate position of the femoral nerve?



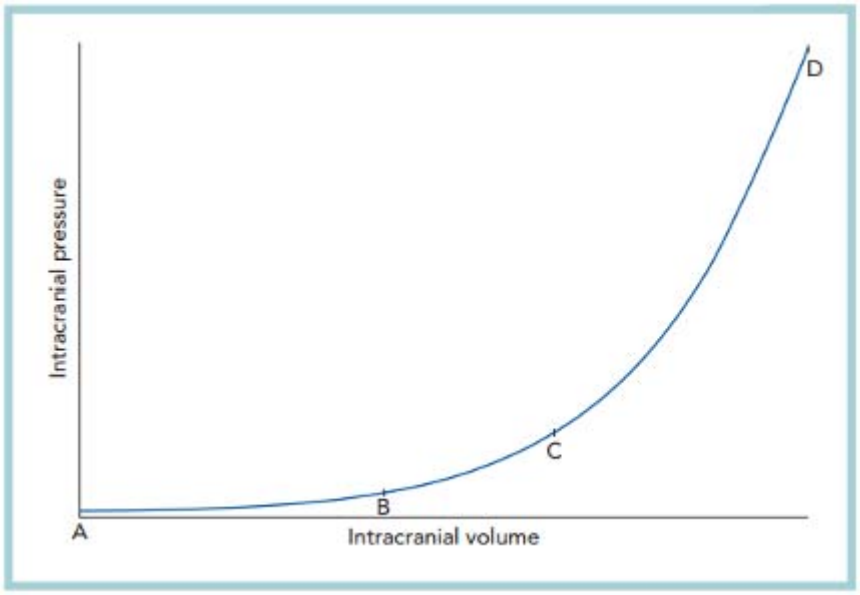
2. An ultrasound image of the groin, enabling accurate identification of the femoral nerve, is shown. The position of the ultrasound probe is shown on the inset. What are the anatomical structures D, E and F?



3. Why are these anatomical structures important, how would you identify them, and why should a neurosurgeon know about them?

Case 9

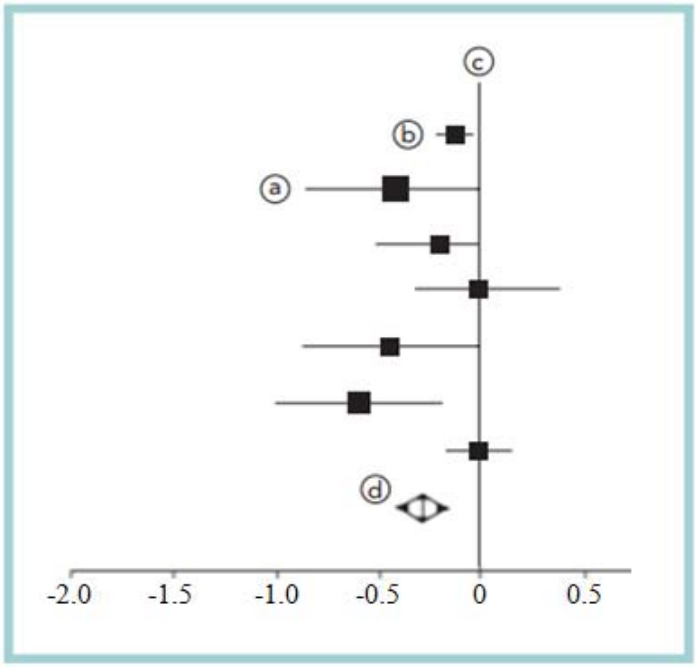
The relationship between intracranial pressure (ICP) and intracranial volume is shown. Consider this graph in the context of a slowly expanding intracranial lesion.



1. Explain what is occurring between points A and B and points C and D.
2. What is normal ICP in an adult?
3. What is cerebral perfusion pressure (CPP)?
4. What influence does ICP have on CPP in a patient with a head injury?
5. What physiological parameters are important in control of CPP?

Case 10

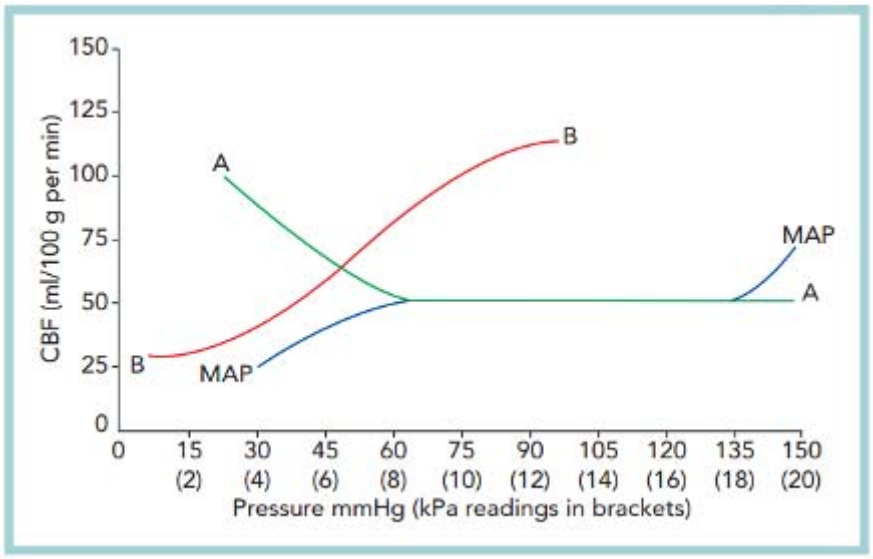
1. What is represented below?



2. Explain the features marked (a) to (d).
3. What are the potential pitfalls of meta-analysis?

Case 11

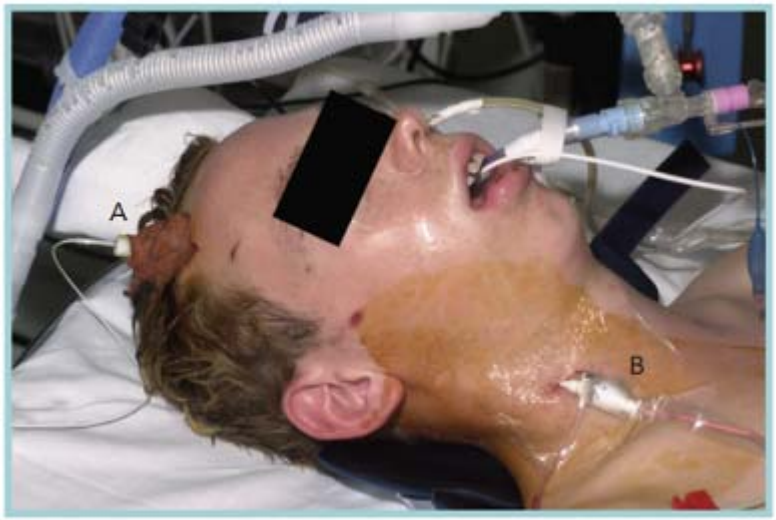
Cerebral blood flow (CBF) on the Y axis against mean arterial pressure (MAP) on the X axis (mmHg values are in brackets) is shown. Two other curves are shown.



1. What does curve A represent?
2. What does curve B represent?
3. What concept does the flat portion between 8 and 18 kPa represent?
4. Explain the concepts illustrated by the graph. How is the graph modified by the effects of underlying hypertension and age?
5. Why is an understanding of this graph essential for the practice of neurosurgery? Are there any other organs within the body that have similar autoregulatory mechanisms, and if so what are they?

Case 12

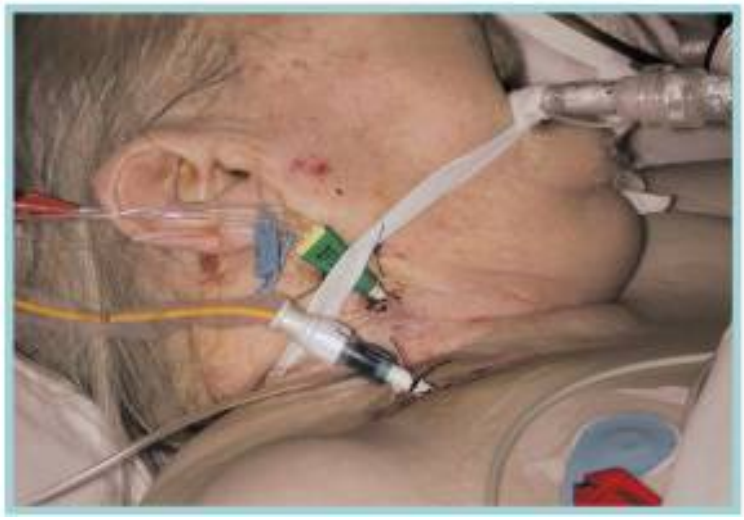
The devices indicated by A and B on this patient are used for monitoring patients with brain injury.



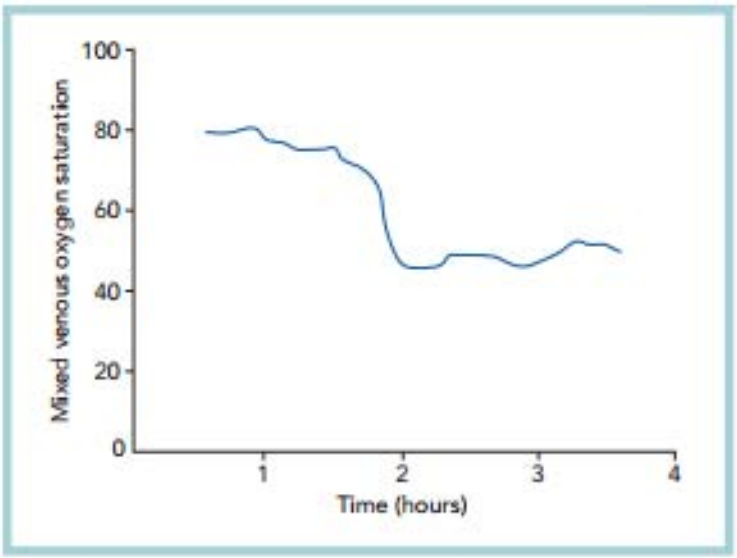
1. What are they?
2. What are the risks associated with their use?
3. Discuss their clinical usefulness.

Case 13

A 69-year-old female with a history of coronary artery disease after a motor vehicle accident has been in the ICU for 12 hours.

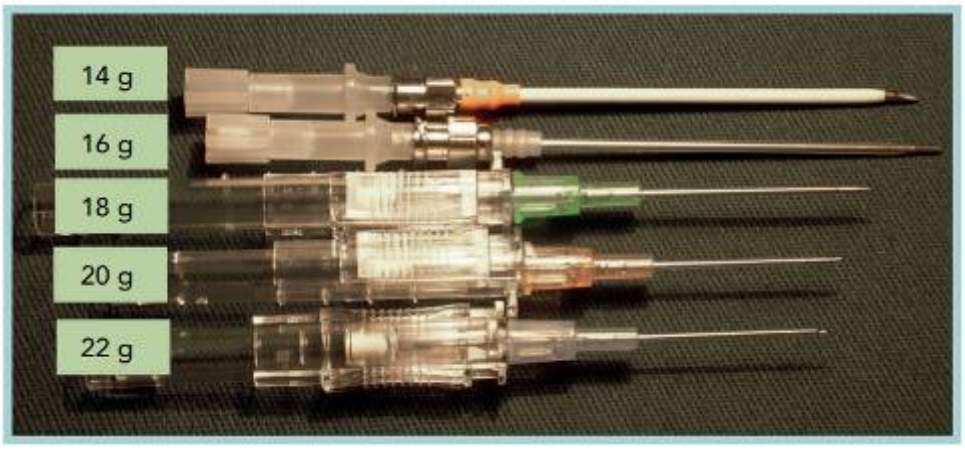


1. What is the yellow catheter contained in a polythene cover and inserted below the green triple lumen CVP line as illustrated?
2. Why is this device used less frequently than it used to be?
3. Why is mixed venous oxygen saturation (MVO_2) a useful parameter to measure in a critically ill patient, and what has this measurement been superseded by?
4. What is the differential diagnosis for the trend over 4 hours in MVO_2 (depicted below) assuming that arterial oxygenation is adequate?



Case 14

An 18 g needle has a greater diameter (is ‘thicker’) than a 22 g needle.

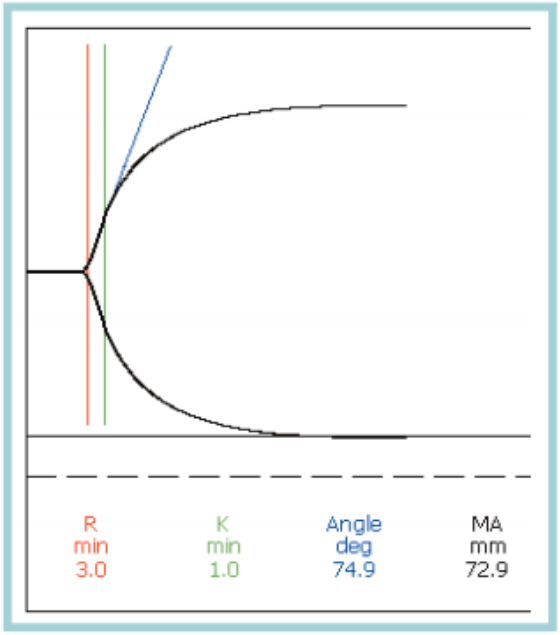


1. What does the abbreviation ‘G’ indicate?
2. What does the abbreviation ‘SWG’ indicate?
3. Why does a higher number (e.g. 22 g) indicate a smaller diameter than 14 g?
4. Does 18 g indicate inner diameter or outer diameter?
5. How long does it take to infuse 1 litre of crystalloid solution through each of the cannulae below, and what is their maximum flow per minute?

- a, 14 g (orange)
- b, 16 g (grey)
- c, 18 g* (green)
- d, 20 g* (pink)
- e, 22 g* (blue)

* = not considered suitable for emergency resuscitation in adults.

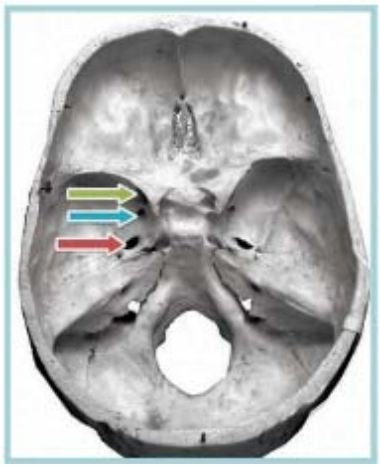
Case 15



1. What is this test called?
2. What is measured by the parameters indicated as R, K and MA?
3. What other tests are available to measure these parameters?
4. Is this a normal test result?

Case 16

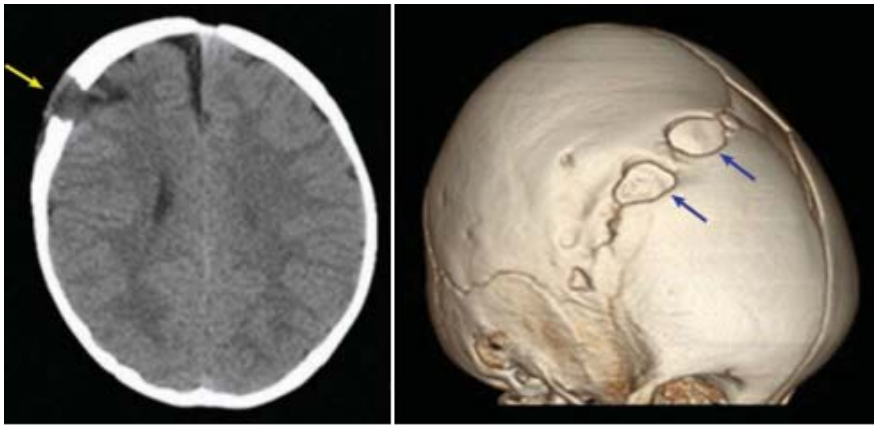
The base of a human skull is shown. The three arrows indicate a different foramen in the skull.



1. What are the three different foraminal indicated by each coloured arrow called?
2. Name the common structure that has branches that pass through each foramen and name each branch.
3. Why is this anatomy important for a neurosurgeon to understand?
4. What practical procedure necessitates the passage of a needle through the foramen marked with the red arrow?

Case 17

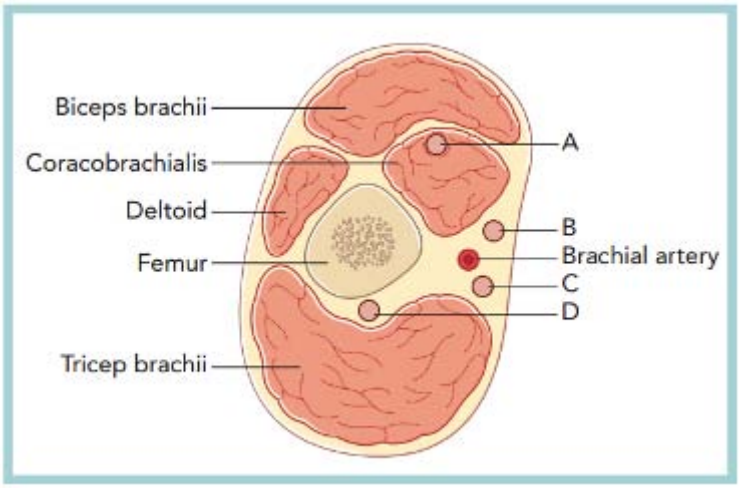
Growing fracture



1. What are the imaging findings?
2. What is your diagnosis?

Case 18

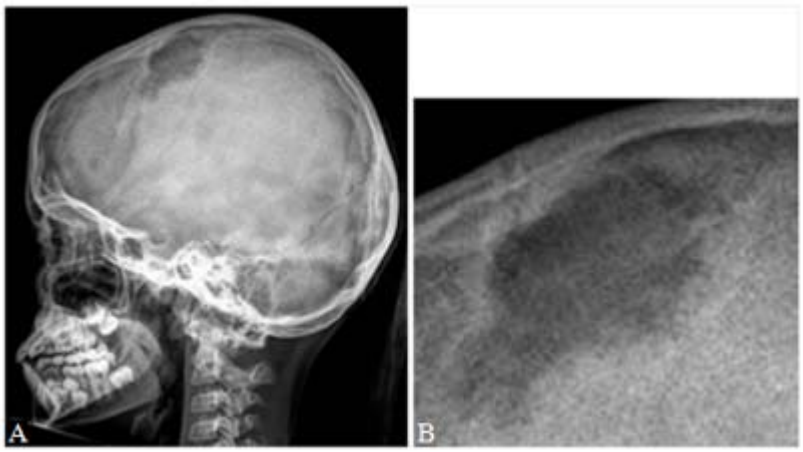
A 52-year-old man with emphysema is scheduled for release of a Dupuytren's contracture in his right hand under regional blockade. He has developed a 'frozen shoulder' and is unable to abduct the arm adequately to perform an axillary block. An alternative block is decided and a cross-sectional view of the arm at the site for this block is shown.



1. What is the name of this alternative regional block?
2. Name each of the nerve labelled.
3. Which of these nerves is commonly missed with the axillary block?
4. Describe the movements you would anticipate on stimulation of each of these nerves.

Case 19

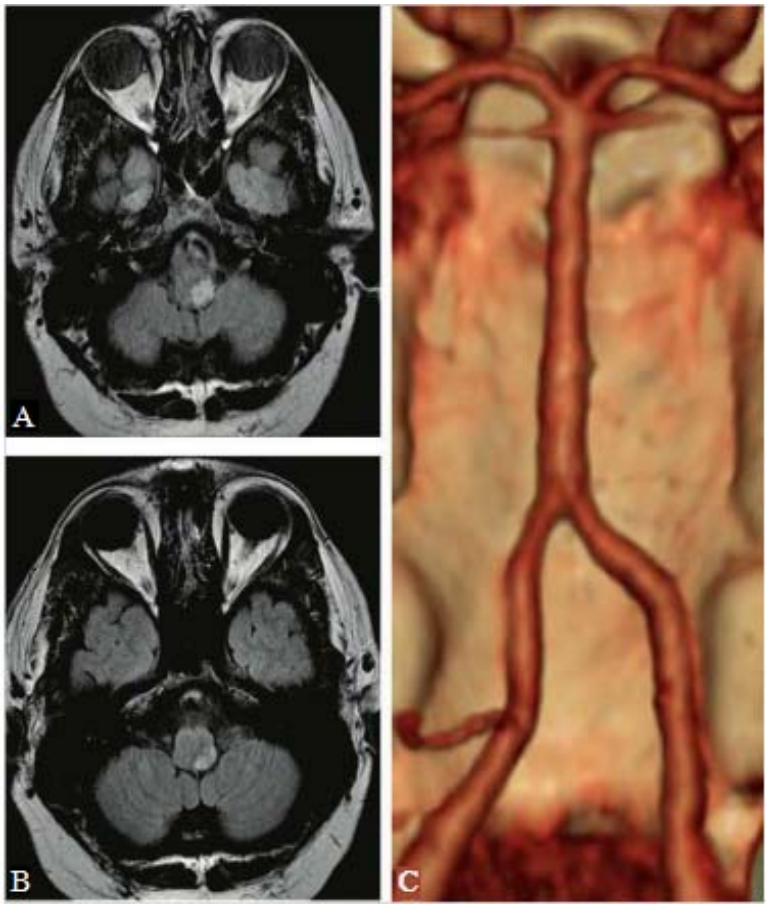
A 9-year-old girl with headaches.



1. What are the imaging findings?
2. What is your diagnosis?
3. What is the differential diagnosis?

Case 20

A 42-year-old man with left facial and contralateral body sensory loss, left-sided ptosis, hoarseness, and vertigo.

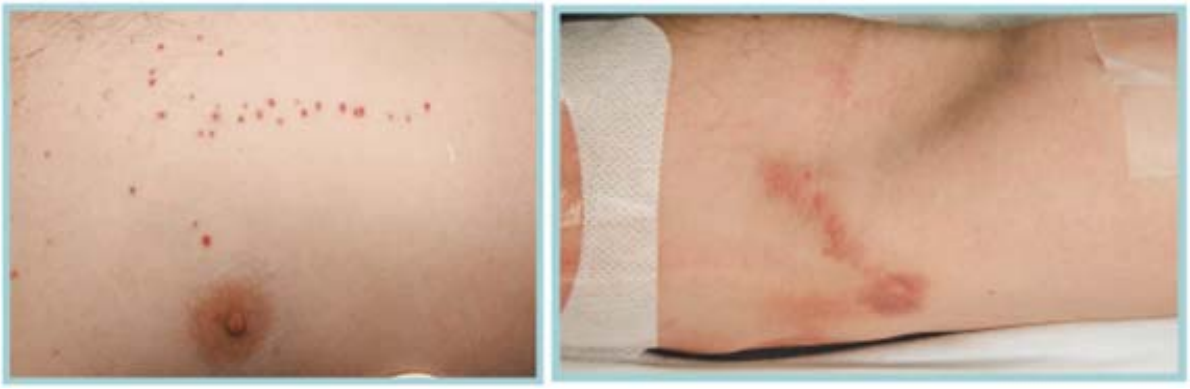


1. What are the imaging findings?
2. What is your diagnosis?

Case 21

An 18-year-old male patient crashed his car. He is behaving inappropriately saying that the car crash was “amazing” and is seemingly oblivious to pain. He has a skull depressed fracture and closed fracture of his right arm.

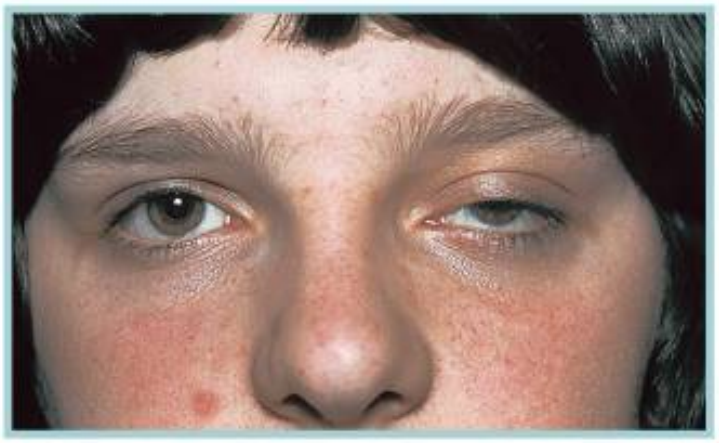
1. List the differential diagnosis that may explain his mental state.
2. Skin lesions noted on his chest (A) and his left antecubital fossa (B) are shown. Do these images help to explain his condition?



3. The neurosurgeon wants to take him to the operating room as soon as possible. Is this patient fit for surgery in his current state? Justify your answer.

Case 22

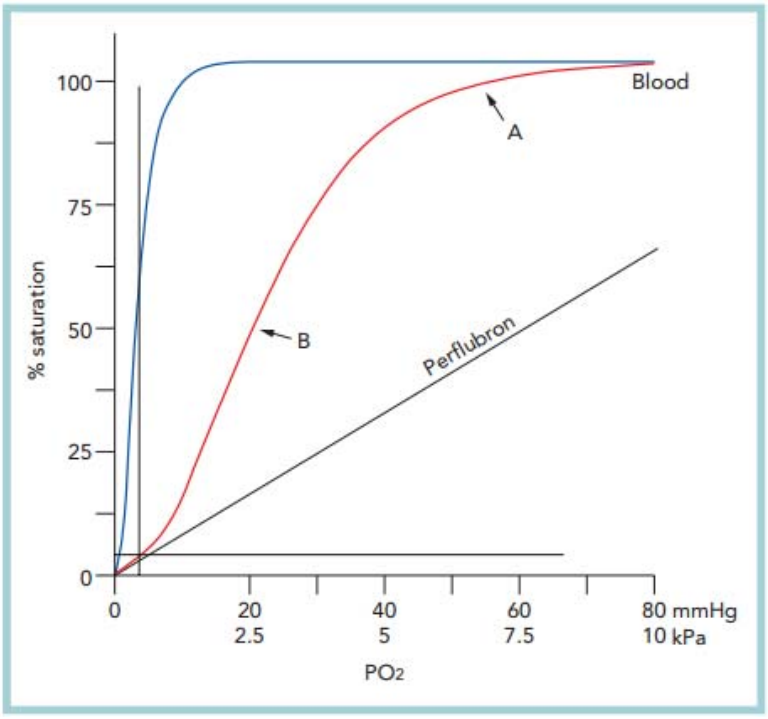
Myasthenia gravis



1. Describe the basic physiological reasons for myasthenia gravis (MG)
2. What are the symptoms of MG?
3. What drug treatments are available?
4. What tests can be employed to aid diagnosis?

Case 23

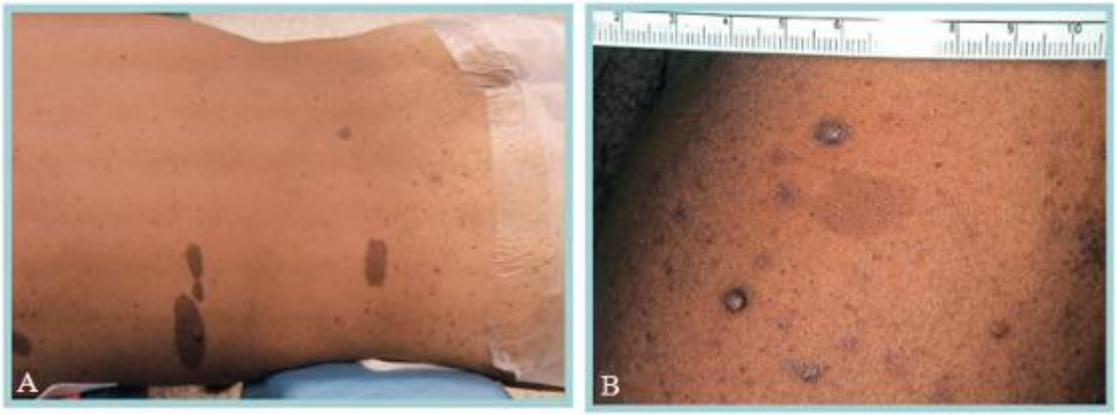
1. What is illustrated in Figure below?



2. What is indicated by the point marked A?
3. What is indicated by the point marked B, and why is it important to understand why this is used?
4. How do the curves of perfluorocarbon solutions, fetal blood and sickle cell blood differ from the normal curve illustrated by the red line?
5. What does the blue line represent?

Case 24

1. What disease is demonstrated in these two images (A, B)?



2. What are these skin lesions called?
3. What tissues are affected by this condition?

Case 25

Consider the figures in the Table below statistically:

DATA TYPE		Measurement scale (nominal, ordinal, interval or ratio)? Parametric or non-parametric?
Female	Male	
Brunette	Blond	
Small	Large	
Cool	Feverish	
Short	Tall	
GCS = 10	GCS = 8	
5 ft	6 ft	
110 lb (50 kg)	220 lb (100 kg)	
37°C (98.6°F)	38°C (100.4°F)	
273° Kelvin	274° Kelvin	

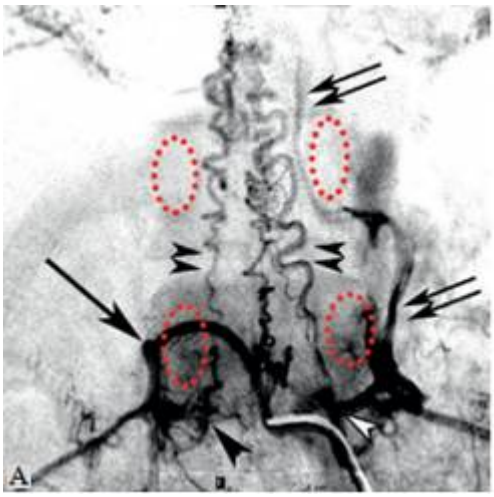
1. Classify from this list each type of data in its appropriate measurement scale: (A) nominal or categorical; (B) ordinal or ranking; (C) interval; (D) ratio or absolute.
2. Are the data parametric or non-parametric?
3. Define each measurement scale listed above (A to D).
4. Which of the four scales are parametric and which are non-parametric?
5. If parametric data are not normally distributed, is it acceptable to use a nonparametric test (e.g. Mann-Whitney U test)?

Case 26

Spinal vascular anatomy and clinical correlation.

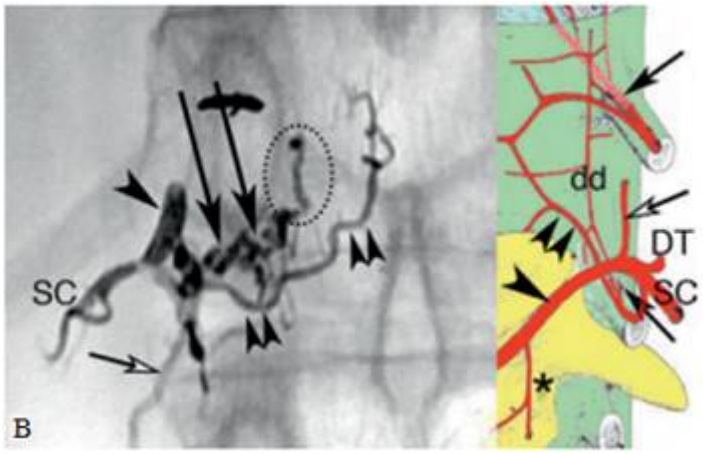
A 64-year-old man presented with progressive, mild, painless, bilateral, lower extremity weakness over the previous 6 months. On examination, bilateral lower extremity power of 4+/5, mild lower extremity hyperreflexia, and reduced sensation up to the midabdomen were present. Magnetic resonance imaging (MRI) of the thoracic spine showed a cluster of intradural, extramedullary serpentine flow voids dorsal to the normal-appearing cord. Time-resolved magnetic resonance angiography (MRA) showed early enhancement of these

enlarged vessels, and a conventional angiogram confirmed a dural arteriovenous fistula (type 1 spinal AVF/dorsal intradural AVF) supplied by the right T12 radicular branch (Fig. shown below, A).



(A) Right T12 segmental artery (*arrow*) courses posteriorly over the right side of the vertebral body to give the dorsal spinal branch under the pedicle (*dotted circle*). Enlarged radicular branches (*arrowhead*) shunt at the level of the nerve root sleeve into the hypertrophied radicular vein ascending through the subarachnoid space to the T10 level, with opacification of the midline anterior and posterior spinal veins as cell as drainage back down through the left T12 radicular vein to exit the spinal canal via intervertebral epidural veins into the inferior hemiazygos vein (*double arrows*).

The fistula was embolized using Onyx 34, which was pushed through the fistulous network and into the intradural draining vein (Fig. shown below, B).



(B) Composite image. Onyx cast outlines the embolized distal segmental artery (*arrowhead*), subcostal artery (SC), radicular branch forming a leash along the nerve root sleeve (*solid arrows*) extending into the radicular vein (*dotted circle*). The pretransverse longitudinal anastomosis (*open arrow*) and dorsal somatic branch (*double arrowheads*) are also outlined. Graphic (yellow vertebra and green dura) also demonstrates the prevertebral anastomosis (*), dural branches (dd), and a radiculomedullary artery at the level above extending into the thecal sac (*solid arrow*).

The patient’s symptoms resolved over 2 months, and follow-up MRI showed no residual intradural abnormality.

Questions

1.Which vascular structures are labeled by the double arrowheads in Fig. A?
A. Anterior spinal artery
B. Posterior spinal artery
C. Radicular veins
D. Paravertebral longitudinal veins
E. Epidural venous plexus

2.What is the most common region supplying the great radiculomedullary artery of Adamkiewicz?
A. Right midthoracic
B. Left midthoracic
C. Right lower thoracic
D. Left lower thoracic
E. Either vertebral artery

3.Which of the following best describes the spinal cord supply from the anterior spinal artery?
A. Centrifugal
B. Centripetal
C. Superficial
D. Pial
E. Circumferential

4.Which of the following is true of the anterior epidural veins?
A. Also known as the anterior extrinsic vertebral plexus
B. Smaller than the posterior epidural veins
C. Regulated by a small number of valves
D. Bidirectional in drainage
E. Arranged in a linear pattern through the epidural space

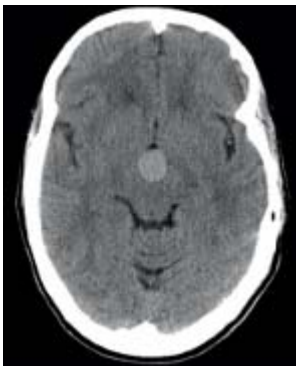
5.Which of the following is true for the sulcocommissural arteries?
A. Arise from the posterior spinal artery
B. Each branch has a territory consisting of the anterior half of the cord
C. Number ~ 400 in total
D. Each branch has a territory consisting of the posterior half of the cord
E. Usually appear as a blush on conventional angiography

6. Which of the following matches of spinal vascular lesion classifications by the traditional Anton-Spetzler and Spetzler modified classification is incorrect?
A. Type I, Intradural dorsal AVF
B. Type II, Intramedullary AVM
C. Type III, Extradural–intradural AVM
D. Type IV, Intradural ventral AVF
E. Type V, Epidural AVF

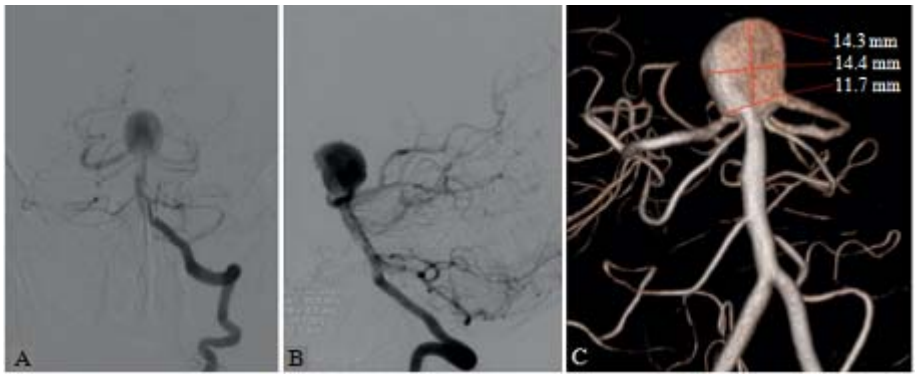
Case 27

Unruptured intracranial aneurysms: endovascular treatment.

A 64-year-old woman presented to her primary care physician after two episodes of "blacking out." Her workup included a computed tomographic (CT) scan of the brain, which revealed a hyperdense lesion in the interpeduncular cistern:



Brain magnetic resonance angiography (MRA) demonstrated a large, basilar tip aneurysm, leading to neurosurgical consultation. Upon examination she was found to be neurologically intact without headaches or an recent syncopal events. Her past medical history was significant for hypertension and tobacco use. She was scheduled for a diagnostic cerebral angiogram under conscious sedation (Fig. below A, B, C) and returned 1 week later to discuss treatment options.



(A) Anteroposterior and (B) lateral digital subtraction angiography showing a basilar bifurcation aneurysm. (C) A three-dimensional angiogram showing a basilar bifurcation aneurysm.

Questions

1. What is the most common method for classifying aneurysm occlusion after endovascular treatment?
A. Toronto Scale
B. Meyers Scale
C. Albany Grade
D. Percent occlusion
E. Montreal Scale

2. What drug is used for anticoagulation when the patient is positive for heparin antibodies?
A. Lovenox
B. Dalteparin
C. Abciximab
D. Argatroban

3. What is the most common cause of mortality during the endovascular treatment of unruptured intracranial aneurysms?
A. Thromboembolic complication
B. Arterial dissection
C. Aneurysm rupture
D. Anesthetic complication

4. What is the most likely cause of morbidity associated with endovascular treatment of unruptured intracranial aneurysms?
A. Thromboembolic complication
B. Arterial dissection
C. Aneurysm rupture
D. Anesthetic complication

5. Approximately what percent volume of the aneurysm should be filled with coils to achieve treatment?
A. 10%
B. 20%

C. 30%
D. 90%
E. 100%

6. Which of the following aneurysms is not approved by the Food and Drug Administration for treatment with a Pipeline Embolization Device (PED)?
A. Large cavernous carotid artery aneurysm
B. Large vertebral artery aneurysm
C. Large petrous carotid aneurysm
D. Large ophthalmic artery aneurysm

7. What is the approximate recurrence rate after endovascular aneurysm treatment?
A. 80%
B. 60%
C. 40%
D. 20%
E. 6%

8. What is the retreatment rate?
A. 40%
B. 20%
C. 10%
D. 5%

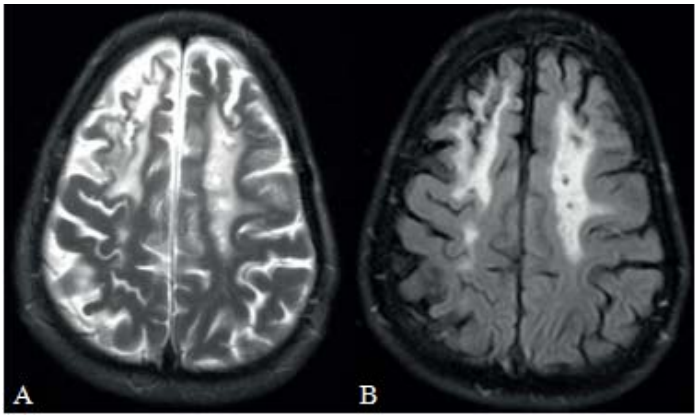
9. What is the single most consistent factor that predicts aneurysm recurrence?
A. Aneurysm neck > 4 mm
B. Dysmorphic aneurysm
C. Large aneurysms (aneurysm diameter)
D. Dissecting aneurysm

Case 28

Moyamoya disease in adults.

A 45-year-old Caucasian woman presents to the emergency department with transient episodes of word-finding difficulty and right arm weakness. She has experienced numerous similar episodes in the past, with increasing frequency over the last 6 months. Her head computed tomographic (CT) scan is unremarkable. She is admitted for further workup. Brain magnetic resonance

imaging (MRI) demonstrates bilateral T2/fluid-attenuated inversion recovery (FLAIR) changes consistent with chronic ischemia, greatest in the watershed zone of the left middle cerebral artery/anterior cerebral artery, and stenoocclusive changes of the internal carotid and middle cerebral arteries bilaterally (Fig. below A, B).



(A, B) Brain magnetic resonance imaging (MRI) demonstrating bilateral T2/fluid-attenuated inversion recovery changes, greatest in the watershed zone of the left middle cerebral artery/anterior cerebral artery, consistent with chronic ischemia.

Questions

1. Which of the following is not true regarding the pathophysiology of moyamoya disease?
A. It is characterized by stenosis or occlusion of the supraclinoid internal carotid arteries.
B. A collateral vascular network is formed through proliferation and hypertrophy of the lenticulostriate arteries.
C. It is associated with other diseases, including sickle cell anemia.
D. It is always unilateral, affecting only one cerebral hemisphere.

2. Which of the following is true regarding the clinical presentation of adult moyamoya disease?
A. Most Asian adults with moyamoya disease present with ischemic symptoms.
B. There is a higher percentage of female patients than male patients.
C. Most North American adults with moyamoya disease present with hemorrhage.
D. The prevalence of the disease is higher in North America than in Asia.

3. Which diagnostic modality is the gold standard for diagnosis of moyamoya disease?

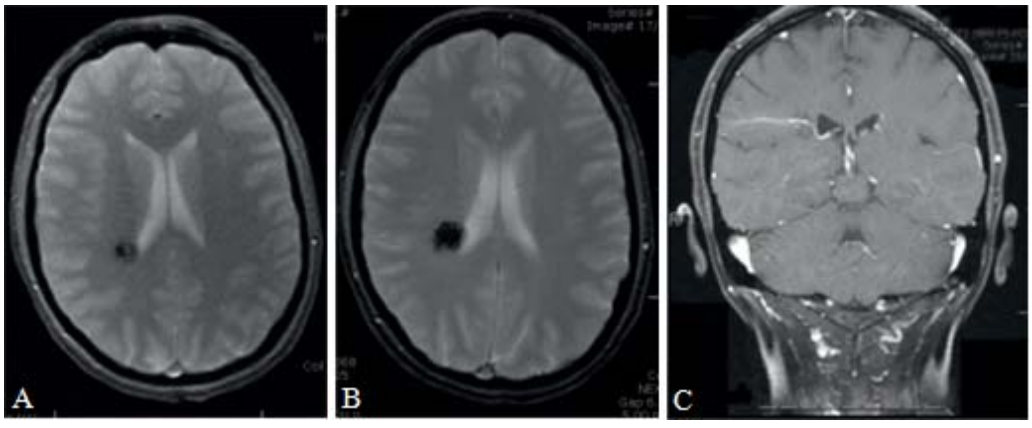
A. Transcranial Doppler ultrasound
B. Noncontrast head CT
C. Digital subtraction angiography
D. Positron emission tomography

4. Which of the following is not a surgical treatment for moyamoya disease?
A. Superficial temporal artery to middle cerebral artery (STA-MCA) bypass
B. Balloon dilatation of the internal carotid arteries
C. Encephalo-duro-arterio-synangiosis (EDAS)
D. Encephalo-duro-arterio-myo-synangiosis (EDAMS)
E. Multiple skull bur holes

Case 29

Cerebral cavernous malformation.

A 45-year-old, right-handed female presented to the clinic with new-onset severe right hemicranial headache, persistent at the right vertex, and associated with subjective left hemibody hypesthesia. The headache has remained refractory to medical management by the neurology team. Five years earlier, brain magnetic resonance imaging (MRI) for unrelated symptoms revealed an incidental deep right parietal hemorrhagic lesion, and no intervention was recommended. Now a repeat MRI scan revealed significant growth of this single lesion (Fig. below A, B).



(A) Axial gradient echo magnetic resonance imaging (MRI) in 2009 revealing a hemorrhagic lesion in the deep right parietal lobe, near the ventricular trigone. (B) Repeat imaging in 2014 after new-onset headaches and sensory symptoms, revealing growth of the lesion. (C) Coronal contrastenhanced T1 MRI revealing a developmental venous anomaly in association with the periventricular lesion.

Questions

1. Which of the following is *true* about familial CCM disease?
A. Familial cases are rare.
B. Familial cases are frequently associated with a DVA.
C. Familial cases affect females predominantly.
D. Familial cases involve one of three known gene loci.
E. Familial cases involve a founder mutation among African Americans.

2. Which of the following is *not* true about imaging features of CCM?
A. Multiple flow voids on T2 MRI sequences with an early draining vein on angiography
B. Faint contrast-enhancement of the lesion proper on T1 MRI sequences
C. Mixed signal appearance with a rim of hypointensity on T2 MRI sequences
D. Multiple hypointense lesions on susceptibility-weighted imaging in cases with family history
E. Faint appearance of the lesion in the late capillary phase of angiography, and frequent prominent vein in the venous phase of the circulation

3. Which of the following is *not* an indication for CCM surgical resection?
A. Recurrent hemorrhage
B. Seizure control
C. Demonstrated lesion growth
D. Established diagnosis in familial cases
E. Neurological deficit

4. Which of the following is *true* about hemorrhage risk from CCM lesions?
A. The risk of overt symptomatic bleeding is the same whether the lesion has bled previously or not.
B. The risk of overt symptomatic bleeding is similar in superficial, deep, and brainstem lesions.
C. A first bleed from a CCM lesion is often fatal.
D. All CCM lesions are associated with some evidence of occult hemorrhage on histology and imaging.
E. An overt symptomatic bleed does not manifest signal features of T1 hyperintensity on MRI.

5. Which of the following is *not* true about CCM and epilepsy?
A. The electroencephalogram (EEG) is always diagnostic in cases of epilepsy associated with CCM lesions.
B. CCM lesions in the temporal lobe are more likely epileptogenic.
C. CCM lesions at the base of the frontal lobe or in the cingulate gyrus may cause partial complex seizures.
D. Lesionectomy is more likely to result in seizure control after recent-onset epilepsy.
E. Resection of epileptogenic tissue beyond the lesion is more likely needed for seizures control in cases with long-standing recalcitrant epilepsy.

6. Which of the following is *true* about CCM treatment?
A. Surgery is always riskier than natural history of the disease.
B. Radiosurgery is a better option that microsurgical resection in the majority of cases with symptomatic CCM.
C. Image guidance is rarely needed for microsurgical resection of CCM.
D. Microsurgical resection should approach a brainstem CCM through a corridor where the lesion presents to a pial or ependymal surface.
E. Microsurgical resection of a brainstem CCM should always include the surrounding hemosiderin-stained brain tissue and an associated DVA.

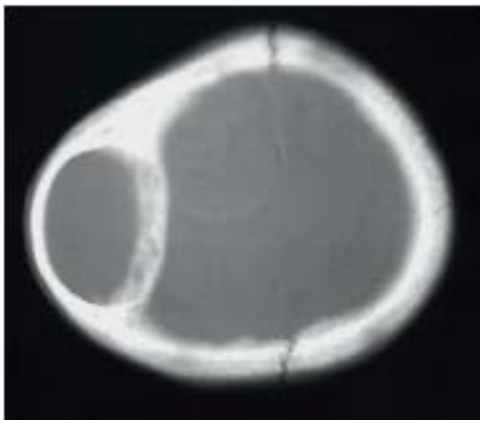
Case 30

Pediatric cephalohematoma.

A 2-day-old male infant presents with an asymmetric scalp mass measuring approximately 3 cm in diameter after a difficult vaginal delivery. The mass is a soft, fluctuant, localized swelling over the right parietal bone, well defined by suture lines.

Presentation of pediatric cephalohematoma at 2 days.

The scalp is freely moving over the swelling, and the child has no neurological deficits. The child was born at term with no history of increased intracranial pressure (ICP) to a nulliparous mother. Delivery was prolonged, with the first stage of labor lasting over 8 hours. The lesion was evaluated, and the decision was made to monitor it carefully over the following few weeks. Over time, the lesion continued to persist and became progressively more firm to palpation. The patient was reevaluated with plain radiography at 7 weeks:



X-ray evaluation of calcified persistent cephalohematoma at 7 weeks

Questions

1. Which of the following aspects of the physical exam is most significant for diagnosis?
A. Size of lesion
B. Depth of lesion
C. Erythema over lesion
D. Definition by suture lines

2. Which factor during delivery does *not* increase the risk of developing cephalohematoma?
A. Prolonged first stage of labor
B. Size of infant relative to birth canal
C. Abnormal presentation of the infant
D. Instrument-assisted delivery

3. Which of the following is a life-threatening condition that must be ruled out during early presentation and requires careful observation?
A. Skin abrasion
B. Caput succedaneum
C. Intracranial hemorrhage

D. Labial tear in mother

4. What is the major indication for surgical intervention?
A. Erythematous changes surrounding the lesion
B. Growth of the lesion to > 6 cm in diameter
C. Ossification of the lesion
D. Fluctuance across suture lines

Answers

Case 1

1. Midline sagittal US images (A) demonstrate a large tubular hypoechoic mass with pronounced vascularity. Axal (B) and sagittal (C) T2 images reveal a large vascular malformation in the region of the vein of Galen, as well as numerous additional enlarged cerebral vessels. The malformation drains into the superior sagittal sinus via a persistent falcine sinus. There is ventriculomegaly with parenchymal volume loss. The sagittal image shows enlargement of the cervical cord and volume loss within the brain stem.

2. Vein of Galen malformation (VOGM).
Vein of Galen malformation (VGAM) is the most common vascular malformation in fetuses and infants. This represents an abnormal communication between intracranial arteries and a persistent embryonic median prosencephalic vein of Markowski (MPV). High flow in the MPV prevents it from regressing to form the normal internal cerebral veins and vein of Galen. Under high pressures, the MPV balloons out and appears as a rounded midline mass with turbulent internal flow, centered in the cistern of velum interpositum and quadrigeminal plate cistern.

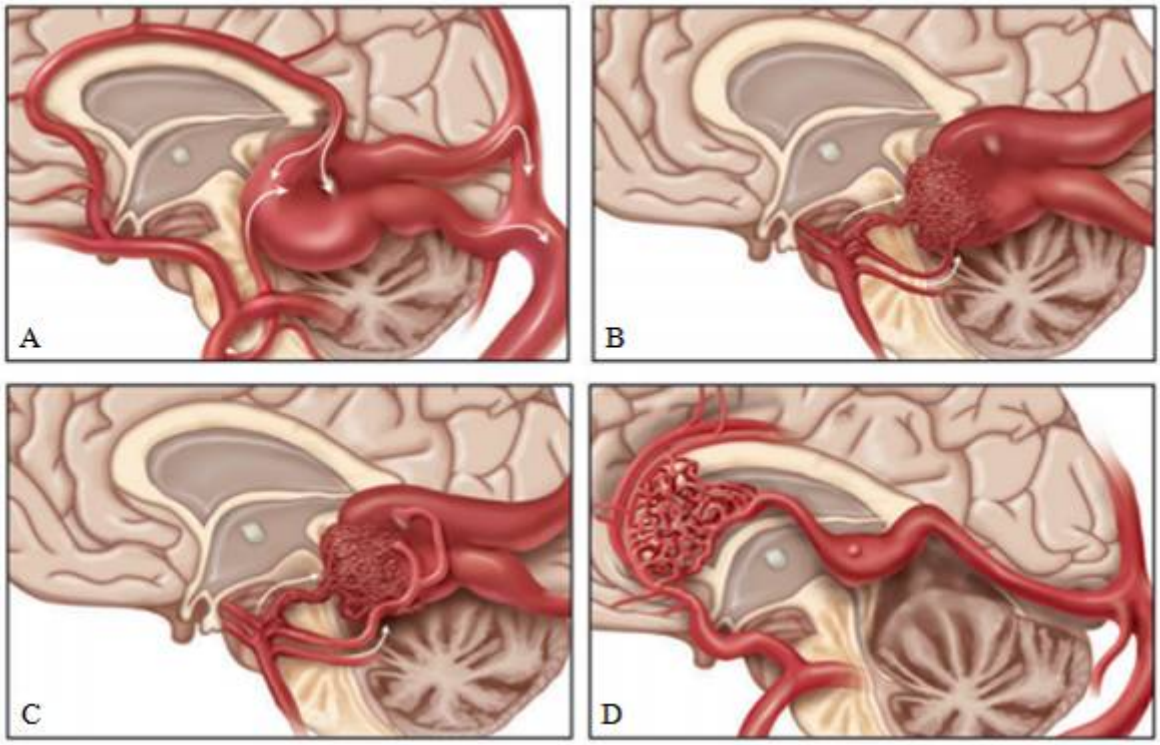
Drainage is into the straight sinus or a persistent embryonic falcine sinus. Severe VGAM can be complicated by hydrocephalus, cerebral ischemia, and high-output cardiac failure. Surrounding tortuous “pseudophlebitic” collaterals are seen in the subarachnoid space. The imaging differential includes vein of Galen aneurysmal dilatation (VGAD), in which a pial or dural AVM drains into a normally formed vein of Galen. Patients present later in life with increased risk of hemorrhage.

3. The most prevalent classification schemes in the literature are those of LasJaunias and Yasargil. The LasJaunias Classification divides VGAMs into two types, mural and choroidal.

Choroidal VGAM: Considered by LasJaunias and colleagues to be a very primitive condition, in which all choroidal arteries contribute flow to the dilated venous pouch, via an intermediate “interposed network” or nidus. This is considered to be the type most often encountered in neonates, presenting with a low Bicêtre score (poor clinical condition).

Mural VGAM: Direct AV fistulous connections into the wall of the MPV. These can be single but are usually multiple. The arteries mal either converge into the MPV, or into separate lobulations of the MPV or occasionally into dilated choroidal venous tributaries to the MPV. This type is thought to be better tolerated and is more often encountered in infants who don’t have cardiac symptoms.

The Yasargil classification divides VGAMs into four categories. The slightly modified version of the classification presented by Pearl and colleagues in 2010 is clinically very helpful:



Modified Yasargil classification of vein of Galen aneurysmal malformation (VGAM).

(A) Type I—one or several direct arteriovenous shunts connected to the median prosencephalic vein (MPV). (B) VGAM type II—a “nidus-like” network interposed between the arterial feeders and recipient venous collector (MPV). Transmesencephalic arterial feeders

are seen**.** (C) Type III—a combination of types I and II. (D) Type IV—not a true VGAM, but rather an arteriovenous malformation draining into an enlarged but normal vein of Galen.
VGAM type I: Direct AV shunts between arterial feeders and MPV. These are typically seen in the neonatal presentation with cardiorespiratory failure.

VGAM type II: With primary subependymal and transmesencephalic posterior circulation supply, with a complex, nidus-like network between the feeding arteries and MPV. This type is more likely to present later in life.

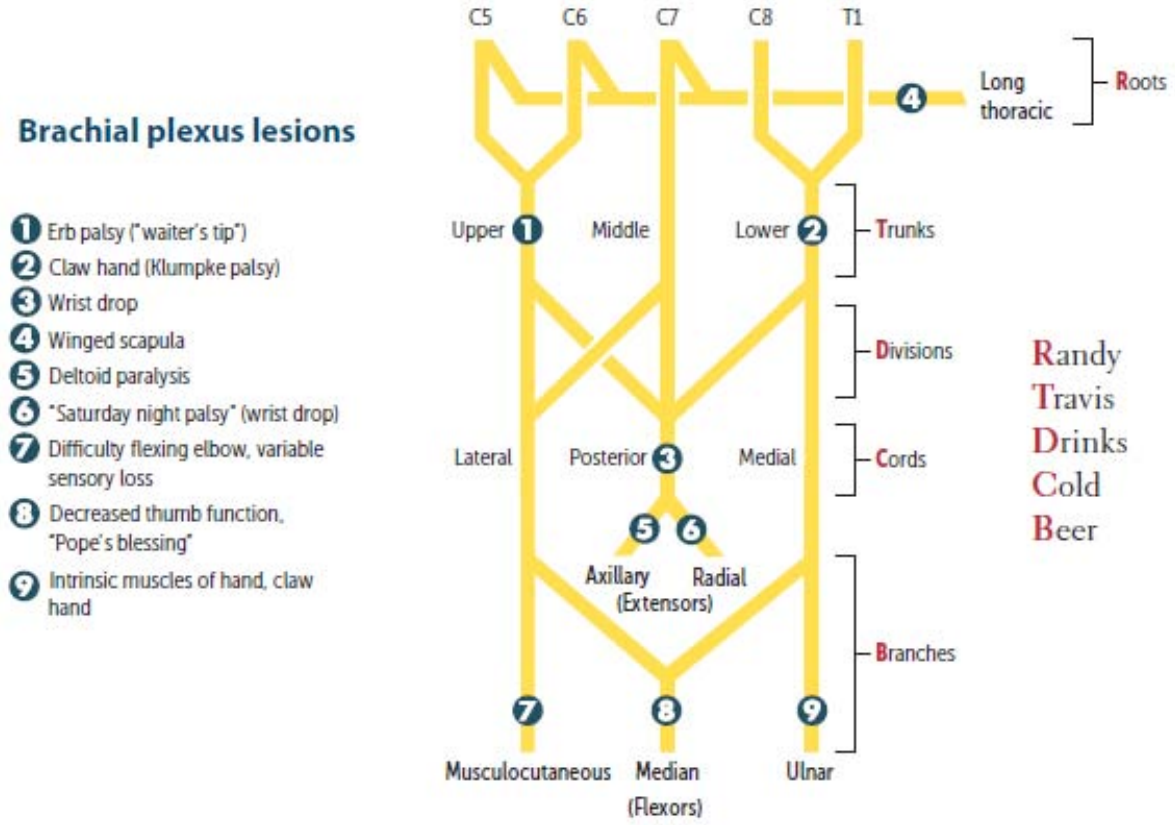
VGAM type III: Combination of types I and II. Type III VGAMS are usually seen in infants or young children.

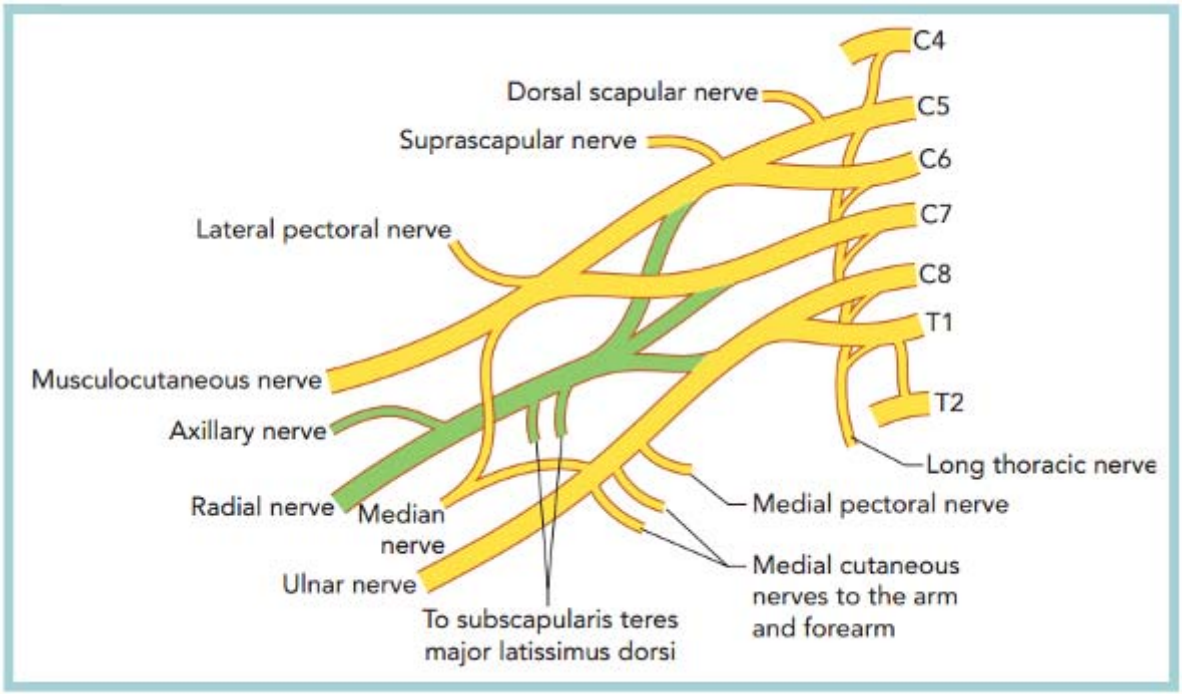
VGAM type IV: AV shunts draining into an embryologically normal vein of Galen, rather than an MPV and thus not true VGAMs. These are classified as vein of Galen aneurysmal dilatation (VGAD) by LasJaunias. Some of these mimic the clinical presentation of VGAMs and are therefore treated similarly.

Despite the presence of these relatively neatly divided classifications, the angioarchitecture of VGAMs is truly more of a spectrum, and not all cases will neatly fit into one of these classifications.

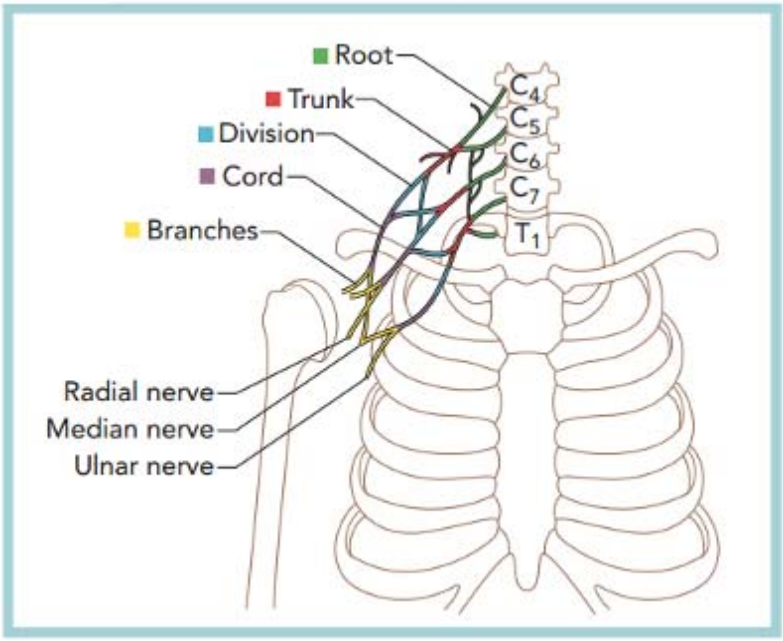
4. Treatment options includes transarterial and/or transvenous embolization, microsurgery, and radiation. Prior to the availability of endovascular embolization, treatment of VOGMs was limited to surgery, which carried a mortality rate of close to 90%. Embolization has significantly decreased mortality and improved clinical outcomes associted with VOGMs. Liquid embolic agents are preferred, and treatmnt is directed to the site of fistulization and feeding arteries.

Case 2





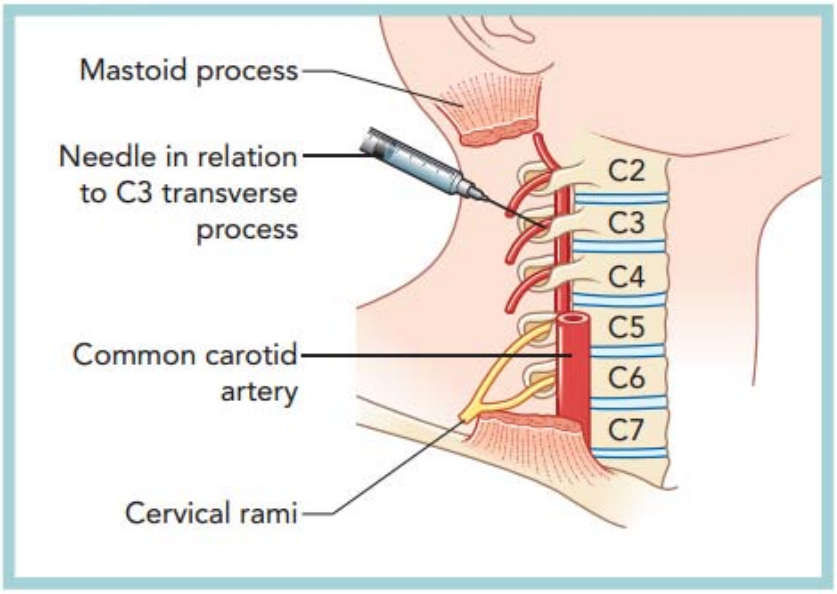
1. Shown in Figures above.
2. Shown in Figure below.



Case 3

1. Yes, but a combination of deep and superficial cervical plexus blocks works best.

2. The block should be performed at C3 (point 3). The mastoid process (point 1) is used to identify the position of the first cervical vertebra.



The cricoid cartilage and the C6 transverse process (Chassaignac's tubercle) are at point 2. Using these two landmarks, the cervical processes are palpated manually. Point 3 is identified approximately 1 cm posterior to the posterior border of the sternocleidomastoid muscle.

3. Dermatomes C2, C3 and C4.

4. The supine patient turns his head to the opposite side from surgery. Under strict sterile technique, intradermal infiltration with local anesthetic is performed and a 2.5 cm 25 gauge needle is introduced at right angles to the skin aiming in a slightly caudal direction (to avoid intrathecal injection). After location of the C3 vertebra (or if the patient reports paresthesia in the distribution of the cervical plexus) 20 ml of local anesthetic (usually 0.375% levo-bupivacaine) is injected. An assistant injects the solution as it is important to keep the needle as still as possible while the injection is taking place. Additional infiltration of the angle of the jaw also prevents the pain of the surgeon's retractor on the angle of the jaw during the procedure. The surgeon may supplement the block by a small amount of local anesthetic on the carotid sheath.

5. A large trial of GA versus LA for carotid endarterectomy demonstrated no difference in the complication rate for either technique.

6. Include: vertebral artery injection producing immediate loss of consciousness or seizure; subarachnoid injection; epidural injection; phrenic nerve palsy, which can lead to serious embarrassment of respiratory function in a patient with advanced lung disease. Less severe complications include local hematoma, transient Horner's syndrome, transient laryngeal nerve palsy and stellate ganglion block.

Case 4

1. The axillary approach to blocking the brachial plexus.

2. Formerly, 'soft' end-points were used to confirm accurate placement of the needle, such as feeling a 'click' on entering the sheath or paresthesia in the distribution of any of the cords of the brachial plexus. These methods can be inaccurate and subjective. Using a current generator and an insulated needle (as shown) to detect paresthesia or contractions/twitching in the hand is the minimum method now expected. The use of ultrasound helps localise the nerve, minimises tissue damage and allows the relationship between the nerve and needle to be directly visualised and subsequent distension of the sheath on injection of the local anesthetic.

Confirmation of successful placement can be made by adequacy of anesthesia or imaging of radiopaque contrast within the plexus sheath. With successful placement it is possible to feel a 'sausage' of local anesthetic within the sheath. The axillary approach, although excellent for forearm and hand surgery, may not anesthetise the musculocutaneous nerve, resulting in tourniquet discomfort, and missing of the eminence of the hand may result.

3. The brachial plexus in the axilla is known to run alongside the brachial artery, so arterial puncture and blood flow is actively sought and then local anesthetic deposited both deep and superficial to the artery. This simple but hazardous technique has been superceded by the use of ultrasound.

Case 5

1. Axial T2 image (A) demonstrates a mixed cystic and solid mass centered within the right thalamus, deep white matter, and medial lentiform nuclei with surrounding vasogenic edema. There is mass effect on the right lateral and third ventricles, midline shift to the left and entrapment of the bilateral atria and occipital horns of the lateral ventricles. T1 postcontrast image with fat suppression (B) reveals solid and ring enhancement. Axial color-coded DTI (C) shows infiltration and displacement of various deep white matter tracts. Sequential reformatted three-dimensional functional images (D) identify the mass with respect to motor tracts for preoperative planning.

2. High-grade tumor centered in deep gray and white matter with diffusion tensor imaging and functional imaging.

Preoperative tractography.

Tractography refers to functinal mapping of tracts, fibers, and interconnections within the brain. lt has a multitude of applications in various clinical settings, including neurodegenerative, demylinating, developmental, psychiatric, and neoplastic proceses. DTI is the basis for tractography and consists of diffusion-weighted imaging (DWI) with directionality.

To obtain DTI maps, a certain minimum number of DWI sequences must be obtained: a *b*0 image and DW images with a *b* value between 700 and 1200 with at least 6 varying motion-probing gradients. In clinical use, 12 or more motion-probing gradients are typically obtained to improve reliability and image quality.

Scan times must be short because patient motion will register as increased diffusivity. Tractography connects fiber orientation within adjacent voxels and infers the overall configuration of white matter. The direction of anisotrpy, and thus the white matter tract orientation, is often color-coded to best identify specific white matter tract on postprocessed images.

Current and potential uses for tractography are vast, ranging from mapping of normal anatomic tracts for preoperative planning to evaluating focal or widespread white matter disease.

Preoperative tractography is useful in both neoplasm and epilepsy tratment planning. With tumors, tractography may be used to evaluate for white matter infiltration or displacement, as well as to map critical white matter pathways with regards to the tumor location to plan the best surgical approach. Similar benefits are noted in epileptic patients with mapping of critical functinal centers and tracts in relation to the seizure focus. Additionally, tractography combined with functional imaging helps identify the locations of language, visual, and motor centers prior to surgical intervention.

Case 6

1. The trigeminal nerve is being blocked, also known as blocking the Gasserian ganglion.

2. It effectively anesthetises the whole of the trigeminal nerve and all its roots. This is a treatment of last resort for severe trigeminal neuralgia. The technique can be carried out with a local anesthetic to evaluate its effectiveness and this can be sufficient to ameliorate the neuralgia. If the block is successful, a permanent block with phenol can be performed, but this is irreversible.

3. The Gasserian gangloin is formed from two roots starting along the ventral surface of the brainstem at the mid-pontine level. The roots pass forward and laterally within the posterior cranial fossa across the superior border of the petrous temporal bone. They enter a recess called Meckel’s cave, which is formed by an invagination of the dura mater of the posterior cranial fossa. In this recess lies the ganglion of the trigeminal nerve and from its anterior border the three divisions of the nerve (ophthalmic, maxillary, mandibular) arise.

The block must be carried out utilising X-ray control and with the head slightly extended. The midpoint zygomatic arch is marked. Approximately 2.5 cm lateral to and slightly above the corner of the mouth on the involved side, a wheal of local anesthetic is raised. A 10 cm 22 gauge needle with stylet in place is inserted so that it will pass through the substance of the cheek, travelling just medial to the ramus of the mandible in a cephalad and medial direction towards the pupil of the eye. The needle tip should then encounter the base of the skull at a point somewhat anterior to the foramen ovale; radiological verification is necessary. The needle is then redirected posteriorly until the foramen is entered. If the patient experiences paresthesia of the mandibular nerve, the needle should be reinserted without paresthesia. One to 3 ml of local anesthetic is injected slowly until the desired clinical effect is reached.

Case 7

1. Axial CT image (A) demonstrates a hyperdense right MCA; normal attenuation is noted within the contralateral MCA and basilar artery. Axial image more superiorly in stroke/ischemia windows (B) shows loss of gray-white matter differentiation involving the right insular ribbon and operculum with mild sulcal effacement. Maximum intensity projection from a CT angiogram (C) reveals occlusion of the proximal-mid right MCA. Axial time to drain (D), cerebral blood flow (E), and blood volume (F) images from a CTP scan in a different patient with similar clinical presentation reveal a central region of significantly decreased cerebral blood flow and increased time to drain with a smaller region of moderately decreased blood volume involving the posterior right MCA vascular territory. This is surrounded by regions of moderately decreased blood flow and increased time to drain with minimal decrease in blood volume.

2. Stroke.

Srtoke is a leading cause of morbidity and mortality, especially in adults. Early recognition and treatment are key to optimizing patient outcomes. Computed tomogrphy (CT) and magnetic resonance imaging (MRI) provide useful diagnostic information, evaluate for potential complications, and aid in directing therapy. Angiography is primarily utilized in cases where intraarterial thrombolysis is a cosideration.

The imaging workup of stroke begins with a noncontrast head CT. CT is useful in identifying early signs of ischemia and evaluating for contrindications to thrombolytic therapy including a large middle cerebral artery (MCA) infarct or the presenc of intrarcranial hemorrhage. Early CT findings of stroke include a hyperdense MCA due to intraluminal clot; loss of gray-white mater differentiation involving the deep gray matter, insular ribbon, and cerebral cortex; and sulcal effacement result from cytotoxic edema and neuronal swelling. In hyper-acute stroke, CT may initially be normal with follow-up imaging hours later better depicting the regions of ischemia. By 24 hours, nearly all stroke are detectable on CT.

MRI is more sensitive than CT, especially within the first few hours of symptom onset. Diffusion sequences may shown restricted diffusion within minutes, manifesting as hyperintensity on diffusion-weighted imaging (DWI) and hypointensity on corresponding apparent diffusion coefficient maps. Development of cytotoxic edema over the next few hours results in increased

T2/fluid-attenuated inversion recovery signal in the involved vascular distribution with loss of gray-white mater differentiation and sulcal effacement in the setting of vessel occlusion, there is loss of the normal T2 flow void in the affected arterial segment. The thrombus typically demonstrates blooming artifact on gradient echo and may appear hyperintense on unenhanced T1 sequences.

3. The advent of intraarterial thrombolysis has greatly improved stroke outcomes. Patients with acute large vessel occlusion (often the proximal MCA) < 5 to 6 hours from symptom onset are potential candidates. Numerous systems are currently available with most combining pharmaceutical and mechanical thrombolytic techniques. Treatment of basilar occlusions may be performed as late as 2 hours after symptom onset in some cases. Potential complications include distal emboli, vessel injury, and parenchymal hemorrhage.

Case 8

1. The femoral nerve should be just lateral to the palpated pulse of the femoral artery (B) pulse and inferior to the inguinal ligament, which runs between the anterior superior iliac spine (A) and the pubic tubercle (C).

2. D = femoral nerve; E = femoral artery; F = femoral vein. Ultrasound is performed to visualise the nerve and an insulated needle used to correctly place a catheter. Paresthesia on insertion of the catheter may confirm correct positioning but is not always detected. Nerve stimulation with electrical current passed through the needle should be used to localise the nerve. The lower the current needed to get contraction of the quadriceps muscles (and in particular movement of the patellar tendon) the higher the success rate of the procedure.

3. The femoral nerve enters the thigh, passing under the inguinal ligament, lateral to the femoral artery. Below the inguinal ligament it divides into anterior and posterior branches. The anterior (superficial) branch supplies sensation to the skin of the anterior and medial thigh and a posterior (deep) branch supplies the periosteum of the femur, the quadriceps muscles, the medial knee joint and the skin on the medial side of the calf and foot (via the saphenous nerve). The block should not be performed lower than the inguinal ligament as one of the branches may be missed. The anatomy of the femoral artery and vein should be known to enable cannulation for intra-arterial pressure monitoring or central venous access, respectively.

Case 9

1. This curve explains the importance of ICP on the injured brain or in the presence of an intracranial space-occupying lesion (e.g. hemorrhage, hematoma, tumor). Between points A and B, a negligible rise in ICP occurs despite increasing intracranial volume. Compensatory mechanisms, namely, displacement of intracranial CSF into the spinal sac, displacement of intracranial venous blood into the extracranial vascular space, reduction of CSF production and increasing CSF absorption all prevent an ICP increase. Between points C and D, a sudden rise in ICP occurs despite only a small increase in intracranial volume. All compensatory mechanisms have been exhausted at this point and ICP rapidly rises. At very high ICP, death from brainstem compression can result.

2. Normal ICP in an adult is 7–15 mmHg in a supine position changing to a negative pressure of up to -10 mmHg in a standing position.

3. CPP = mean arterial pressure (MAP) – ICP.

4. Normally CPP is equivalent to MAP as ICP is low (–10 to +10 mmHg). With intracranial pathology, ICP may rise to 15–20 mmHg. Above 25 mmHg active measures should be taken to reduce ICP, as CPP will be reduced. CPP should not be allowed to fall below 70mmHg, as below this level secondary insults to the injured brain start to occur.

5. Hypoxia and hypercarbia must be prevented in brain injured patients as both increase cerebral blood flow and, therefore, volume. MAP should be maintained at a level of at least 90 mmHg (assuming that the ICP is <20 mmHg), as this will maintain the critical CPP of 70 mmHg.

Case 10

1. This is a ‘Forest Plot’ or ‘blobbogram’, which is a diagrammatic representation of a meta-analysis of various randomised controlled clinical trials. This example is a meta-analysis of seven randomised controlled trials.

2.
(a) Each *horizontal line* represents the randomized comparison of individual trials. The width of each line represents the 95 per cent confidence interval (CI). If the CI crosses the vertical line, it means there is no true difference between the two groups in the study.

(b) Each black *square* represents the point estimate of the effect. This is the most likely value and the results become less likely as they move towards the 95 per cent CI limits. The *size of the square* is proportional to the number of the subjects in the study.

(c) The *vertical line* represents the "line of no effect" – depending on the positive or negative effect of intervention, the blobs are placed to the right or left of this line. If the variables are dichotomous, the line of effect would correspond to 1.

(d) The *diamond* is the weighted average of all the studies. It provides a point estimate with 95 per cent CI. In this case, the diamond does not cross the line of no effect, hence is statistically significant, although the effect size is less than 0.5.

3. Meta-analysis can give misleading results. The most common problems are:
• Publication bias: positive studies are more likely to be published than negative studies, skewing the estimate of effectiveness of a treatment.
• Methodological weaknesses of individual trials, particularly smaller trials, weaken a meta-analysis.
• Heterogeneity of studies: subtle differences in study design, methods or patient population can cause pooled data to be inaccurate.

Case 11

1. Curve A shows the relationship between arterial O_2 tension and CBF. O_2 only influences CBF if pO_2 is <8 kPa. Avoiding hypoxia prevents secondary brain insult after head trauma but has little effect on CBF at normal O_2 levels. Excess (100%) O_2 can reduce CBF by up to 10%.

2. Curve B shows the linear relationship between CBF and arterial CO_2 between values of 3 and 10 kPa. This is important when intracranial space-occupying lesions (SOLs) (e.g. head injury, cerebral edema, other brain pathology) cause limited intracerebral compliance. A low normal $PaCO_2$ (between 4.0 and 4.6 kPa [30 and 35 mmHg]) temporarily reduces CBF, preventing brain swelling and increased ICP. Prolonged mild hyperventilation, however, alters cerebrospinal bicarbonate and CBF increases again 6–12 hours after any change of pCO_2.

3. It shows cerebral autoregulation. CBF, at 45–60 ml/100g brain tissue/min, is maintained at a constant level between MAPs of 60 and 30 mmHg. (CBF grey matter approx. 80 ml/100g/min; CBF white matter approx. 20 ml/100g/min). Outside this range, flow varies passively with pressure. Without autoregulation, brain perfusion is susceptible to sudden fluctuations in arterial BP caused by changes in posture and/or environmental stresses.

4. Hypertension and increasing age cause a long-term change in the relationship between BP and CBF, shifting the curve to the right.

5. Cerebral and renal autoregulation are comparable but renal blood flow (RBF) is regulated more by the microcirculation (i.e. glomerular afferent and efferent arterioles). RBF and CBF, when compared across physiological BPs, are very similar.

Case 12

1. A is an intracranial pressure (ICP) monitor, also known as an ICP bolt. The illustration shows an intraparenchymal fibreoptic transducer. Other types are available. B is a jugular bulb catheter.

2. The risks of both devices are infection and hemorrhage. Experienced personnel should insert these devices under full sterile technique.

3. In head injury, ICP monitoring is important for both recognition of a sudden increase in ICP secondary to an acute bleed and also for calculation of cerebral perfusion pressure (CPP). CPP is compromised secondary to any intracranial mass within the vault of the skull (e.g. a hematoma, a cerebral contusion or generalised cerebral edema secondary to a diffuse brain injury). A CPP above 70 mmHg is a goal of head injury management and is calculated thus:
CPP = MAP (mean arterial pressure) – ICP.
CPP guides the neurosurgeon/intensivist as to when to manipulate MAP with fluids and vasopressors.

A jugular vein catheter measures oxygenation in venous blood leaving the brain by a fibreoptic transducer connected to a light emitting diode and works by the same principles as a pulse oximeter. It is placed within the jugular sinus by being passed retrogradely up the internal jugular vein. Normal saturation of jugular venous blood ($SjVO_2$) is 55–75%.

Low $SjVO_2$ implies excessive O_2 extraction by brain tissue and a sluggish cerebral blood flow (CBF). Therapeutic manoeuvres include checking for good oxygenation and correct positioning of the patient's head to assist adequate arterial flow and venous drainage. Echocardiography, a pulmonary artery catheter or at least a CVP line should be used to ensure adequate fluid loading and allow safe use of vasopressors (e.g. noradrenaline) to elevate MAP to optimise cerebral perfusion.

A high $SjVO_2$ may be due to either excess CBF (luxury perfusion) or a lack of metabolic activity within cerebral tissue because of cell death. CT can distinguish between these two situations. With 'luxury perfusion', high ICP and high $SjVO_2$ may require mild hyperventilation to a pCO_2 of 4–4.5 kPa (30–35 mmHg) to reduce CBF by vasoconstriction. Once PCO_2 is optimised, if $SjVO_2$ remains high, sedatives can be added to suppress metabolic activity. Although the catheter picks up global cerebral ischemia, focal ischemia may not be detected and caution should be taken not to overinterpret a normal or high jugular venous oxygen level.

Case 13

1. A pulmonary artery (PA) catheter is inserted into the right internal jugular vein. PA catheters were frequently inserted into critically ill patients because by having their tip in the PA they enabled both cardiac output (CO) and MVO_2 to be measured. These two measurements allow calculation of oxygen delivery and consumption.

2. Other less invasive ways of assessing CO are now available. PA catheter insertion can lead to tricuspid/pulmonary valve damage, arrhythmias and pulmonary infarction. TOE and/or less invasive CO measurement (e.g. Lidco, Picco) have replaced the PA catheter. MVO_2 measurements have been largely superseded by central venous oxygen saturation measurement, which is a close approximation of MVO_2 and only requires sampling from a central venous line. Similar conclusions can be drawn from both saturations measurements.

3. MVO_2 is the percentage oxygen saturation in venous blood from the body as a whole and is measured accurately from samples taken through the pulmonary artery catheter. MVO_2 is the end result of blood leaving tissues after they have consumed oxygen and represents a balance between arterial oxygen delivery and tissue oxygen consumption.

Oxygen delivery = CO × oxygen content (Hb × % saturation × 1.34)
This is helpful in critically ill patients because if they are shocked or septic, insufficient tissue perfusion is occurring and MVO_2 falls. Measures can then be taken to boost oxygen delivery (e.g. checking arterial oxygen is sufficient, fluid loading to try and increase CO, inotropic support and/or increasing Hb level, so called ‘goal-directed therapy’, the goal being a MVO_2 of 70% or greater.

4. If arterial hypoxia is not present, oxygen delivery must be failing due to decreased CO from either hypovolemia or ventricular dysfunction leading to inadequate oxygen delivery to the tissues. Increased oxygen consumption from shivering and/or fever or sepsis can also increase oxygen uptake, causing a reduction in MVO_2.

Case 14

1. The abbreviation ‘g’ indicates gauge (i.e. the diameter of the needle).

2. It indicates Standard Wire Gauge. Earlier names associated with this form of gauge are, in the UK, Holtzapffel and Stubs Wire Gauge. In the USA the terms US Birmingham Wire Gage and British Standard Wire Gauge were used.

3. Historically, during the wire making process, a rod of metal is heated and ‘pulled’ (stretched) through a specific sized hole in a draw plate. With each additional pull through the next smaller hole in the draw plate, the wire is stretched to a thinner gauge. The SWG indicates the number of pulls through the machine. Hence, a 22 SWG has been pulled through a smaller hole than a 16 gauge. Each pull (and decrement in size) had to be within specific tolerances, so that the wire would not break during the pulling process.

4. It indicates the outer diameter.

5. Minimum times for 1 litre infusion and maximum flow rates in ml/minute are:
a = 3.7 minutes (270 ml/minute);
b = 4.2 minutes (236 ml/minute);
c = 10 minutes (103 ml/minute);
d = 15 minutes (67 ml/minute);
e = 32 minutes (31 ml/minute).

Case 15

1. A thromboelastogram (TEG); also known as a thrombelastogram.

2. R is the clotting time, which indicates the time from the start of the test until clotting starts. This time is influenced by platelet function and clotting factors. K is the angle or rate of clot formation, which is influenced by platelet function, clotting factors and fibrinogen concentration. MA is the maximum amplitude, which measures the strength of the clot and is influenced by platelet function and fibrinogen concentration.

3. Skin ‘bleeding time’ was used extensively, but has been found to have a poor sensitivity and a poor specificity for clotting and platelet function. The platelet function assay measures the function of platelets and can be used to determine the residual effects of aspirin-like compounds on the clotting cascade in a patient.

4. It is a normal test result.

Case 16

1. Green arrow = superior orbital fissure; blue arrow = foramen rotundum; red arrow = foramen ovale.
2. The trigeminal nerve, which has three branches: ophthalmic, maxillary and mandibular. Each branch passes through the respective foramen as listed above.

3. The anatomy of the foramen ovale must be known in order to use regional anesthesia for the treatment of intractable trigeminal neuralgia pain.
4. The entire trigeminal nerve can be blocked by injecting local anesthetic through the foramen ovale under radiological control.

Case 17

1. Axial CT shows a distracted right frontal bone fracture with transcalvarial herniation of leptomeninges (arrow) and underlying encephalomalacia.
3D CT reconstruction shows scalloped erosions (arrows) along the fracture margins.

2. Leptomeningeal cyst.

Compared to the adult skull, the pediatric skull is thin and incompletely ossified, causing it to be more fragile as well as more elastic. Since the dura mater is more adherent in children, skull fractures carry an increased risk of dural tear and underlying brain injury. In distracted fractures, the arachnoid and pia mater can herniate out through the defect to produce a leptomeningeal cyst. This interposed tissue inhibits bone healing, and CSF pulsations cause progressive bone erosion, colloquially known as a “growing fracture.” At imaging, the margins of the fracture are angular and scalloped, with an intervening encephalocele. The underlying brain is encephalomalacic and may also herniate into the fracture (traumatic meningoencephalocele).

Case 18

1. A mid-humeral block.

2. A = musculocutaneous nerve; B = median nerve; C = ulnar nerve, D = radial nerve.

3. The musculocutaneous nerve. It emerges from the brachial plexus above the usual puncture site for an axillary nerve.

4. The mid-humeral block is performed at the level of the insertion of deltoid muscle. Each nerve can be identified individually with a nerve stimulator:
• Musculocutaneous nerve: flexion of the elbow, supination of the forearm.
• Median nerve: pronation of the forearm, flexion of the lateral three fingers,

flexion and adduction of the thumb; flexion and abduction of the wrist radially.
• Ulnar nerve: flexion and abduction of the wrist in an ulnar direction, flexion of the medial two fingers.
• Radial nerve: extension of the elbow, extension and abduction of the wrist radially, extension of the fingers.

Case 19

1. Lateral radiograph of the skull (A) demonstrates a solitary lytic lesion with lobulated margins and well-defined borders involving the parietal bone just posterior to the coronal suture. Coned-down view (B) reveals a "beveled edge" appearance without sclerotic margins.

2. Langerhans cell histiocytosis (LCH).

Langerhans cell histiocytosis, previously known as histiocytosis X, is a proliferative disorder of Langerhans cells in the bone marrow, skin, and/or lymph nodes. Unifocal LCH (eosinophilic granuloma) affects the skeletal and/or pulmonary systems. Multifocal unisystem LCH (Hand-Schüller-Christian disease) is characterized by exophthalmos, diabetes insipidus, and lytic calvarial lesions.

Multifocal multisystem LCH (Letterer-Siwe disease) is an acute disseminated disease affecting multiple organs. Bone involvement is lytic with ill-defined margins and periosteal reaction in the acute phase, and well-defined with sclerotic margins in the chronic phase. Asymmetric erosion of the inner and outer tables of the skull produces a “beveled edge” sign in tangential views and a “bullseye” appearance en face. There may be a central fragment of devascularized bone, known as a “button sequestrum.” Multiple confluent lesions may produce a “geographic” skull, with large lytic areas surrounded by normal bone. Lesions within the maxilla and mandible can result in “floating teeth.” Other lytic skull" lesions include dermoid/epidermoid cyst, hemangioma, fibrous dysplasia, early Paget disease, hyperparathyroidism, multiple myeloma, metastasis, and infection. Clinical history and time course aid in diagnosis.

3.
• Epidermoid cyst.

Epidermoid cysts result from abnormal deposition of epithelial rests within the diploic space during development. They are a relatively common cause of solitary lytic skull lesions in the pediatric population and tend to occur along

sutural lines. When an epidermoid cyst involves the diploic spaœ, the radiographical appearance may overlap that of LCH. Epidermoid cysts are generally well-defined expansile lesions without a central matrix. They may or may not have a sclerotic rim. On MRI, epidermoid cysts are iso- to hypointense to cerebrospinal fluid (CSF) on T1 and iso- to hyperintense on T2 with characteristic increased signal on diffusion-weighted imaging.

• Malignancy.
The most comon malignancies to involve the calvarium in children include lymphoma, leukemia, Ewing sarcoma, and metastatic neuroblastoma. Rhabdomyosarcoma may also invide the calvarium via direct spread. Malignancies often result in bony expansion and destruction, which manifests as poorly defined lucencies. Aggressive periosteal reaction may be noted, especially with Ewing sarcoma. Lesions may be solitary or multiple; multiple lesions are suggestive of hematogenous spread. On MRI, malignancies demonstrate marrow infiltration or bony expansion and desruction with a soft tissue mass. The soft tissue component is typically hypointense on T1 and hyperintese on T2 with avid but heterogenous enhancement.

Case 20

1. Axial FLAR images (A, B) demonstrates abnormal increased signal intensity within the posterolateral medulla on the left. Three-dimensional volume rendered image from a computed tomography angiogram (CTA) (C) reveals absence of the left PICA; a portion of the proximal right PICA is visualized on the contralateral side. Although visualization of PICA on CTA is variable, occlusion was confirmed on conventional angiogram (not shown).

2. Lateral medullar syndrome (LMS).

LMS or Wallenberg syndrome bassically results from occlusion of the posterior inferior cerebellar artery (PICA) with associated infarct of the posterolateral medulla. Similar findings may be seen with involvement of the vertebral artery as well. The majority of cases occur in older patients secondary to thromboembolic disease; in yonger patints, vertebral artery dissection associated with trauma or vasculopathies are the most common etiologies.

Clinically, patients present acutely with loss of pain and temperature sensation involving the ipsilateral face and contrlateal body, ipsilateral Horner syndrome, ipsilateal limb ataxia, and vertigo. Hoarseness and difficulty swallowing

comonly occur secondary to cranial nerve involvement. The clinical symptoms and prognosis correlate well with the overall size of the infarct; patients with larger infarcts have a worse prognosis. The majority of patients, however, have a relatively good outcome with improvement of symptoms over time.

Computed tomography is often normal due to the inherent difficulty in evaluating posterior fossa structures. Magnetic resonance imaging (MRI) is more sensitive, demonstrating regions of increased T2/FLAIR signal along the posterolateral medulla.

lsolated medullary lesions may be subtle and only visualized on the inferior-most axial images. Larger infarcts may also involve the inferior cerebellum and are more conspicuous. Restricted diffusion and mass effect/edema is seen acutely. Subtle enhancement may be seen in the subacute phase. Chronically, there is volume loss of the involved segment.

Case 21

1. The patient has mania, which can result from a number of causes namely: bipolar disorder; stimulant drug abuse (e.g. cocaine, amphetamines); medications (e.g. steroids or selective serotonin reuptake inhibitors); non-compliance with medications for bipolar or psychotic disorders; thyrotoxicosis; intracranial malignancy.

2. The lesions on the chest are known as skin popping lesions and are caused by injecting drugs intradermally. In this case the patient had injected cocaine into his chest skin. The lesion in his antecubital fossa shows where he has attempted to inject into his brachial vein. These two signs indicate that substance abuse is highly likely to be the cause for his mania.

3. Surgery for a depressed fracture is urgent but the effects of cocaine should wear off within 15 minutes if the drug was injected or within an hour if there are still intradermal deposits. Cocaine can cause severe hyperthermia, hypertension, arrhythmias and convulsions, especially in overdose, so it is worth waiting for the acute effects to wear off. A history should then be taken to establish an alternative diagnosis as listed above. Assuming the mania was simply due to the cocaine, surgery can then proceed. If mania recurs in the postoperative period, an extended recovery with sedation, ventilation and ICU admission may be needed.

Case 22

1. MG is a disorder of neuromuscular transmission caused by either genetic anatomical or acquired immunological abnormalities of the neuromuscular junction (NMJ). Anatomical distortion of the postsynaptic muscle membrane, or its disruption by antibodies, reduces the effect of acetylcholine (ACh) and decreases nerve impulses.

2. MG is often picked up by ocular signs (ptosis or diplopia – often unilateral) (66% of cases) or oropharyngeal muscle weakness (difficulty chewing, swallowing or talking) (16%), but limb weakness is rare. Symptoms worsen as the day progresses or with fatigue, emotional upset, systemic illness (especially viral infections), hypothyroidism or hyperthyroidism, pregnancy, the menstrual cycle, drugs affecting neuromuscular transmission and increases in body temperature. After 15–20 years the weakness may become fixed, with atrophic muscles.

3. There is no complete treatment for MG. Cholinesterase inhibitors (e.g. pyridostigmine/neostigmine) retard enzymatic hydrolysis of ACh at cholinergic synapses, allowing ACh accumulation at the NMJ and prolonging its effect. Marked improvement can occur with steroids. Azothiaprine, cyclosporine or cyclophosphamide are second-line treatments but may cause immunosuppression. Plasma exchange and IV immunoglobulin can alleviate symptoms for short-term benefit prior to surgery.

4. The Tensilon® test assists diagnosis. Weakness improves after IV edrophonium chloride. IM neostigmine, which has a longer duration of action, may work better in some patients and is particularly useful in children, whose response to IV edrophonium chloride may be too brief to observe. Electromyography and repetitive nerve stimulation (RNS) are useful for diagnosis. A significant decrement to RNS in either a hand or shoulder muscle is found in most (60%) patients with MG.

Case 23

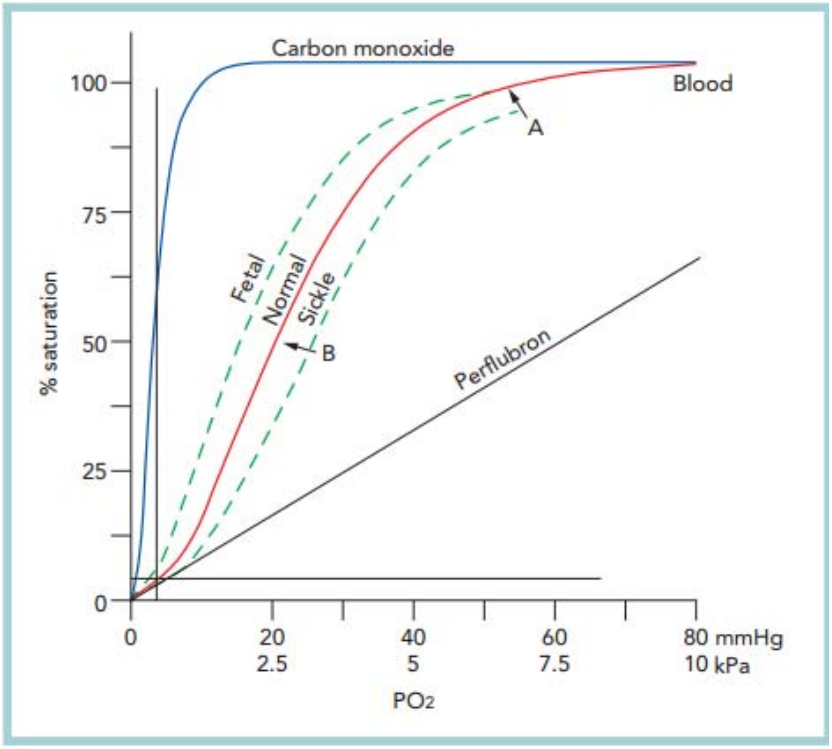
1. The oxygen dissociation curve for normal adult hemoglobin (Hb).

2. This point indicates an Hb O_2 saturation of 97%, above which good oxygenation of blood is guaranteed; this is the norm for an adult breathing room air at sea level.

3. The P50 (normal value 3.73 kPa (28 mmHg)). This is the O_2 partial pressure in blood that leads to a Hb O_2 saturation of 50%. P50 is used to compare O_2 carriage in normal Hb with other types of Hb such as fetal Hb, abnormal variants of the Hb such as methemoglobin, sickle cell anemia, thalassemia and other abnormal Hbs.

4. Perfluorocarbons do not have a sigmoid shaped dissociation curve, so oxygen carriage is linear. Perfluorocarbons need a high inspired O_2 concentration before clinically useful O_2 carriage can occur. Fetal Hb is designed to function at lower O_2 tensions than adults and therefore is to the left of the adult curve. The sickle cell Hb O_2 dissociation curve is to the right of the normal curve and loads O_2 less avidly than normal Hb. Sickle cell Hb is poorly soluble and crystallises, causing red cell deformation (sickling) with red cell fragility and thrombus formation.

5. CO has an affinity with Hb 240 times more than O_2. In CO poisoning the dissociation curve of carboxyhemoglobin is very much shifted to the left (blue line on Figure below). Carboxyhemoglobin is incapable of O_2 carriage, so high apparent 'saturations' actually reflect poor available O_2.



Case 24

1. This patient has Von Recklinghausen's neurofibromatosis (neurofibromatosis 1).

2. Image (A) shows flat uniformly pale brown macules, known as Café-au-lait spots, which vary in size from 0.5 to 20 cm and can be found on any cutaneous surface. Six or more spots greater than 1.5 cm in diameter are presumptive evidence of neurofibromatosis in children over 6 years of age and adults. The raised lesions in (B) are neurofibromas.

3. The lesions in neurofibromatosis affect tissues derived from neuroectoderm. These include neurofibromas, plexiform neurofibromas, optic nerve gliomas and astrocytomas of the brain and spinal cord. Hamartomas and meningiomas may also develop. The disease is genetic and autosomal dominant, although manifestations may not occur until early childhood or adulthood.

Case 25

1. The classification is shown below.

2.

DATA TYPE		Measurement scale (nominal, ordinal, interval or ratio)? Parametric or non-parametric?
Female	Male	Nominal, non-parametric
Brunette	Blond	Nominal, non-parametric
Small	Large	Ordinal, non-parametric
Cool	Feverish	Ordinal, non-parametric
Short	Tall	Ordinal, non-parametric
GCS = 10	GCS = 8	Ordinal, non-parametric
5 ft	6 ft	Ratio, parametric
110 lb (50 kg)	220 lb (100 kg)	Ratio, parametric
37°C (98.6°F)	38°C (100.4°F)	Interval, parametric
273° Kelvin	274° Kelvin	Ratio, Parametric

3. (A) Nominal or categorical scales are groups of data that do not bear any mathematical relationship to one another and are all 'equal' categories (e.g. one eye color is not 'larger' or 'better' or ranked any differently from another eye color).

(B) Ordinal or ranking scales (ranked from large to small, or small to large) are groups of data points that bear a mathematical relationship to one another (e.g. one subject is sicker, more traumatized, more dehydrated than another).
(C) Interval scales are measurements where the intervals (distances) between data points are fixed, linear and constant over the whole range of measurement. The size of the difference between 10 and 11 kg is the same as the difference between 100 and 101 kg.
(D) Absolute or ratio scales are data with an absolute zero point (e.g. lbs or kg weight). Temperature in degrees Kelvin has an absolute zero point, so it is an absolute scale. Temperatures in Celsius/Fahrenheit, have arbitrary zero points in each scale (0° Celsius is freezing, and 0° Fahrenheit is 32°F below freezing point). The term 'ratio scale' is also used because weight scales (ounces, kg) have true zero points. The ratio between any two points is independent of the unit of measurement (e.g. the ratio of 20 kg to 10 kg is 20/10 = 2). The ratio between the same mass in lbs is 44 lb/22 lb, also 2. The ratio of body temperature to room temperature in °C and °F are not the same. 37/20°C is 1.85 while in °F 98.4/60 is 1.64. Temperatures measured in °C and °F are interval not ratio scales.

4. Categorical and ordinal scales are non-parametric.
5. Yes. 'Downgrading' data that requires fewer assumptions is acceptable but if data are downgraded, less information from the data set is used, so a statistical difference is more difficult to detect. Skewed data may be missed.

Case 26

1. C. Radicular veins
2. D. Left lower thoracic
3. A. Centrifugal
4. D. Bidirectional in drainage
5. C. Number ~ 400 in total
6. E. Type V, Epidural AVF

Comment

A thorough knowledge of spinal vascular anatomy is necessary for correct diagnosis and appropriate and safe management of vascular spinal lesions, including dural arteriovenous fistulas (Anton-Spetzler [AS] type I), cord AVMs (AS type II), metameric AVMs (AS type III), pial AVFs (AS type III), epidural AVFs, spinal aneurysms, vertebral neoplasms (aggressive hemangiomas,

vascular metastases, aneurysmal bone cysts), and vascular cord neoplasms, such as hemangioblastomas. The segmental organization of the spinal vasculature is consequent to the early development of the 31 separate metameric units constituting the somites (vertebrae, soft tissues), neural crests (nerve roots, dorsal body), and adjacent neural tube (spinal cord).

Case 27

1. E. Montreal Scale
2. D. Argatroban
3. C. Aneurysm rupture
4. A. Thromboembolic complication
5. C. 30%
6. B. Large vertebral artery aneurysm
7. D. 20%
8. C. 10%
9. C. Large aneurysms (aneurysm diameter)

Case 28

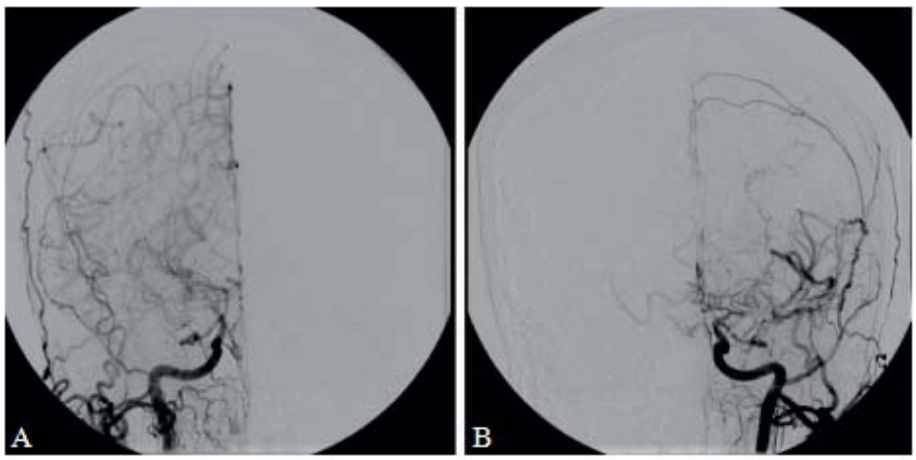
1. D. It is always unilateral, affecting only one cerebral hemisphere.
2. B. There is a higher percentage of female patients than male patients.
3. C. Digital subtraction angiography
4. B. Balloon dilatation of the internal carotid arteries

Comment

Moyamoya disease (MMD) is an idiopathic cerebrovascular disorder characterized by stenosis or occlusion of the supraclinoid internal carotid arteries, with subsequent hypertrophy and proliferation of lenticulostriate arteries to form a collateral network.

The name stems from the appearance of these collaterals on conventional catheter angiography—*moyamoya* translated from Japanese means “a hazy cloud like a puff of cigarette smoke.” *Moyamoya syndrome or phenomenon* refers to a similar condition that occurs when an underlying etiology, such as atherosclerosis, radiation therapy, or sickle cell disease, produces unilateral or bilateral steno-occlusive arterial changes and associated moyamoya collaterals.

Digital subtraction angiography (DSA) has been the gold standard in the evaluation and diagnosis of MMD from the time of its initial description (Fig. below A, B).



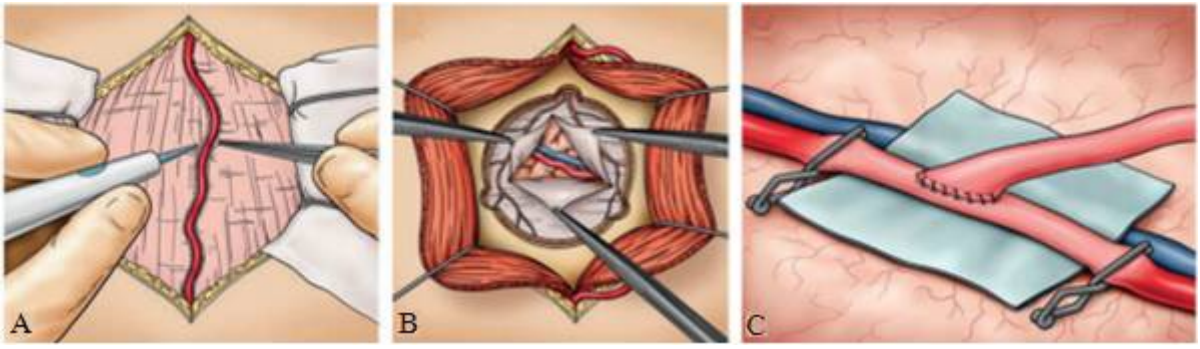
(A, B) Changes of moyamoya disease with occlusion of the right internal carotid artery terminus, marked narrowing of the left A1 and M1 segments, and occlusion of the left M1 prior to its bifurcation. Flow to the left anterior middle cerebral territory is via a markedly enlarged middle meningeal artery, which supplies transdural collaterals.

Suzuki described six separate angiographic stages of MMD based on the pattern of steno-occlusion and collateral formation. Grade I refers to narrowing of the internal carotid artery (ICA) apex without moyamoya collaterals. Grade II includes ICA stenosis along with initiation of moyamoya collaterals. Grade III is progression of the ICA stenosis with intensification of moyamoya collaterals. Grade IV refers to development of external carotid artery (ECA) collaterals. Grade V refers to intensification of ECA collaterals along with reduction of moyamoya collaterals. Grade VI represents the final stage of the disease process, with total occlusion of the ICA and disappearance of moyamoya collaterals. Though the Suzuki grading scheme is commonly used to categorize the severity of MMD in patients, how each grade affects the natural history and response to treatment is poorly understood.

For years, surgical revascularization has been the mainstay of treatment for patients with MMD. For patients with ischemic symptoms, hemodynamic disturbances rather than thromboembolic events are felt to be the principal underlying etiology; therefore, a variety of surgical procedures have been devised to augment cerebral blood flow (CBF) distal to the area of steno-occlusion and decrease the risk of further ischemic events. For patients with hemorrhagic symptoms, similar surgical procedures have been employed in the hope that augmentation of CBF will ultimately lead to a reduction in the fragile moyamoya collaterals and a decrease in further hemorrhagic events. These procedures fall into two categories:

(1) direct revascularization, where a scalp artery or other extracranial vessel is sewn directly to a cortical artery in an effort to provide an immediate increase in CBF; and (2) indirect revascularization, where vascularized tissues are applied to the cortical surface in an effort to promote angiogenesis and improved CBF over time.

The most common direct revascularization procedure for MMD is the superficial temporal artery to middle cerebral artery (STA-MCA) bypass (Fig. below A, B, C).

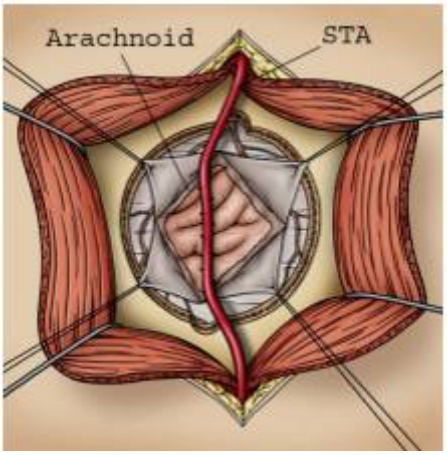


Superficial temporal artery to middle cerebral artery (STA-MCA) bypass. (A) After opening of the skin, the STA is dissected free along its length with a small 5 mm cuff using either sharp dissection or needle cautery. (B) A craniotomy is performed with multiple bur holes to preserve the middle meningeal artery, and the dura is opened to find a recipient branch of the MCA. Note that the STA is positioned safely behind the temporalis muscle. (C) The anastomosis is performed in a running fashion with the 10–0 monofilament nylon sutures used to initially anchor the donor vessel. Alternatively, multiple interrupted sutures can be used for the anastomosis (not pictured).

Briefly, the course of the STA is mapped with a Doppler flow probe, and the posterior limb of the STA is usually selected. The STA is dissected free, along with a small cuff of galea connective tissue, and a small craniotomy centered over the distal aspect of the sylvian fissure is performed. The dura is opened and an M4 branch of the MCA is selected. The donor STA and recipient M4 branch are then temporarily clipped, prepared, and anastomosed in end-to-side fashion using 10–0 interrupted or running sutures. Temporary clips are removed, and patency of the graft is confirmed using intraoperative catheter angiography or, more recently, indocyanine green (ICG) videoangiography and/or perivascular flow probe.

The most common indirect procedures includes encephalo-duro-arterio-synangiosis (EDAS), encephalo-duro-arterio-myo-synangiosis (EDAMS), and multiple bur holes. All are designed to promote the formation of extracranial to

intracranial collaterals over a period of months to ultimately achieve revascularization of the cerebral hemisphere.



Encephalo-duro-arterio-synangiosis. This procedure involves suturing a branch of the superficial temporal artery with its associated cuff of galea connective tissue to the pia surface of the cerebral cortex.

For EDAS (Fig. above), the STA and accompanying cuff of galea connective tissue are exposed in a manner that preserves STA inflow and outflow. This arterialized tissue is then overlaid and sutured onto the brain surface. For EDAMS, the STA and accompanying cuff of galea connective tissue are overlaid and suture onto the brain surface in conjunction with a pedicle of temporalis muscle. For multiple bur holes, 10 to 20 bur holes are placed over each affected hemisphere, and the underlying dura is incised and separated. Although predominantly used in children, this technique has been reported with successful results in adults as well.

The primary goal of surgical revascularization for MMD is to decrease the incidence of future ischemic and hemorrhagic events.

Case 29

1. D. Familial cases involve one of three known gene loci.
2. A. Multiple flow voids on T2 MRI sequences with an early draining vein on angiography
3. D. Established diagnosis in familial cases
4. D. All CCM lesions are associated with some evidence of occult hemorrhage on histology and imaging.
5. A. The electroencephalogram (EEG) is always diagnostic in cases of epilepsy associated with CCM lesions.

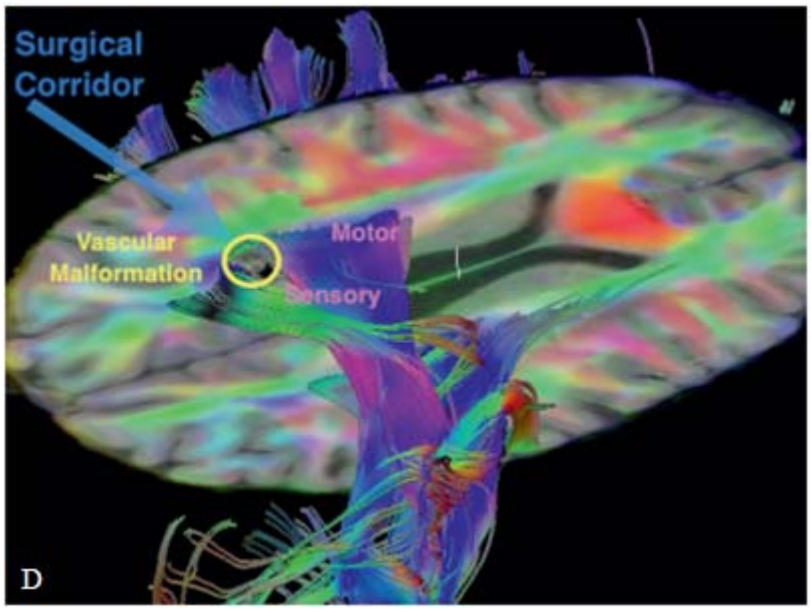
6. D. Microsurgical resection should approach a brainstem CCM through a corridor where the lesion presents to a pial or ependymal surface.

Comment

Signal features included popcorn mixed intensities on T1 and T2 sequences and a surrounding hemosiderin ring. Gradient echo sequences revealed a blooming effect within the lesion, consistent with chronic blood products. There was faint contrast enhancement in the hemorrhagic lesion proper. Immediately adjacent and anterior to it, there was a contrastenhancing vessel with transmedullary course and caput medusae appearance, consistent with developmental venous anomaly (DVA, Fig. C). There were no other lesions elsewhere in the brain on gradient echo and susceptibility sequences.

Diffrential diagnosis, clinical decisions, and surgical course

The lesion presented characteristic imaging features of cerebral cavernous malformation (CCM), also known as cavernous angioma or cavernoma. The growth of the lesion raised a differential diagnosis of neoplasm, but the absence of lesional contrast enhancement or surrounding edema, and the presence of an associated DVA, made this differential unlikely. The possibility of an arteriovenous malformation (AVM) was considered, but the popcorn appearance and hemosiderin ring and the absence of associated arterial feeders or lesional contrast enhancement made this diagnosis unlikely.



(D) Functional MRI with tensor diffusion imaging of sensorimotor fiber tracts in relation to the lesion. The surgical corridor is chosen to avoid disruption of these tracts.

The solitary nature of the lesion (absence of additional lesions elsewhere in the brain on gradient echo and susceptibility sequences) and the associated DVA essentially excluded the familial autosomal dominant form of CCM disease. The lesion had demonstrated significant hemorrhagic growth during expectant followup, associated with new severe localizing headache. Consideration of surgery was recommended in view of demonstrated lesion growth and new headaches. Functional MRI allowed mapping of sensorimotor cortical areas, and tensor diffusion imaging highlighted respective fiber projections. The lesion abutted just posterior to the capsular sensory fibres projecting to the postcentral gyrus and well posterior to motor fibers projecting from the precentral gyrus (Fig. above D).

A surgical corridor was chosen through the superior parietal lobule, minimizing disruption of these subcortical tracts. Image-guided navigation was performed along with monitoring of somatosensory evoked response. Stimulation of white mater fibers during surgery ensured that the dissection did not venture near motor fibers.

The CCM was successfully resected, separating it and preserving the associated DVA. Surgical appearance was that of mulberry, and histopathological features included dilated sinusoidal capillary spaces, lacking mature vessel wall angioarchitecture, and associated hemosiderin-stained brain, all typical of CCM. The somatosensory evoked responses from the left median but not tibial stimulation became slightly attenuated during the course of the resection. The patient awoke with mild loss of light touch sensation and proprioception in the left leg, but with preserved motor function and tone, and visual field. The sensory loss improved gradually in the few weeks after the procedure.

Case 30

1. D. Definition by suture lines
2. A. Prolonged first stage of labor
3. C. Intracranial hemorrhage
4. C. Ossification of the lesion

Comment

Pediatric cephalohematoma is a collection of subperiosteal blood, typically located on the parietal or occipital region of the calvarium. The condition is

named for its occurrence in neonates; shearing forceps on the scalp and skull sustained during labor cause a separation between the skull and periosteum and subsequent rupture of blood vessels. Rarely, cases of cephalohematoma occur in juveniles or adults after trauma or surgery. In infants, the hematoma is usually observed within the first 1 to 3 days of birth. The majority of cases spontaneously resorb within the first month of life, and children typically have no neurological deficit. Potential causes for this condition includes a rigid birth canal, relatively large size of the neonate compared with the birth canal, abnormal presentation of the fetus, and delivery assisted by an instrument.

Indications for surgery

Calcification or ossification is rare but may occurs in cases that do not resolve. X-ray or computed tomographic (CT) scan of the skull is indicated in patients whose hematoma has not resorbed within 6 weeks after birth. There is much ebate regarding conservative treatment (e.g., observation) versus an early surgical approach to cephalohematoma because there are no controlled trials for reference. Percutaneous aspiration of the lesion increases the risk of infection and is contraindicated except for evaluation of potential infection. The majority of calcified cephalohematomas will still resolve in 3 to 6 months and are treated conservatively according to guidelines.

In patients with later-stage calcification or ossification, advocates for surgery argue that outcomes are much better with intervention at an earlier are due to the natural molding process and decreased risk of raised intracranial pressure, in addition to cosmetic advantages. Ossified cephalohematoma has hardened rather than resorbed, and it has a clearly defined inner and outer layer of bone surrounding the lesion. The inner layer is composed of the inner and outer table of the infant’s intramembranous calvarial bone, and the outer table consists of subpericranial bone derived from the separated pericranium. The contour of the inner lamella can either follow the convex shape of the skull or become concave, encroaching upon the cranial vault space.

The surgical approach involves removal of the overlying newly formed bone, the soft tissue mass, and the underlying original bone. Multiple techniques for bone replacement have been used, each showing favorable and uneventful outcomes in case studies. After removal, the depressed underlying region is often sectioned into multiple pieces and remodeled as a bone graft, with good results and no residual evidence in the patient after a few years.

References

1. Robert E. Harbaugh, Christopher Shaffey, William T. Couldwell, Mitchel S. Berger. Neurosurgery Knowledge Update. A Comprehensive Review. Thieme Medical Publishers, Inc., New York, 2015.

2. Robin Bhatia, Ian Sabin. Challenging Concepts in Neurosurgery. Cases with Expert Commentary. Oxford University Press, Oxford, UK, 2015.

3. Magnus A. Garrioch, W. Bosseau Murray. Anaesthesia. Illustrated Clinical Cases. CRC Press, New York, 2015.

4. Thankamma Ajithkumar, Adrian Harnett, Tom Roques. Oxford Case Histories in Oncology. Oxford University Press, Oxford, UK, 2014.

5. Graeme Mcleod, Colin McCartney, Tony Wildsmith. Principles and Practice of Regional Anaesthesia, 4th Ed., Oxford University Press, Oxford, UK, 2012.