Headaache :

Hallmark of Neurological Emergencies In Pregnant and Post-partum Women

By

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Aim

- Diagnostic evaluation of common and serious neurological emergencies that present in pregnant and post-partum women.

- To help us and obstetricians to avoid misdiagnosis in high-risk patients.

Importance

- If specific treatments not started promptly, result in morbidity or mortality in these young previously healthy individuals.

- If a patient has poor outcome, the medical, social, and medico-legal impact is often high.

Pathophysiology

- Unique pathophysiological states of pregnancy and puerperium
- Raised oestrogen conc stimulate production of clotting factors, increases risk of thromboembolism
- Increased plasma and total blood volumes increase risk of hypertension
- Raised progesterone concentrations enhance venous distensibility, potential leakage from small blood vessels.
- High oestrogen levels fall in postpartum period.
- *Combined, these hormonal changes result in leaky capillaries and vasogenic oedema*

Introduction

Acute neurological symptoms in pregnant and post partum women

- exacerbation of pre-existing neurological condition
- initial presentation of non-pregnancy-related problem
- new acute-onset neurological problem

Pregnant and postpartum patients with headache and neurological symptoms diagnosed with

- pre- eclampsia;
- cerebral venous sinus thrombosis
- reversible cerebral vasoconstriction syndrome.

Pre-eclampsia

- *new onset of hypertension and proteinuria after 20 weeks in a previously normotensive woman.*
- *Criteria for severe pre-eclampsia are two occasions of hypertension (blood pressure greater than or equal to 160/110 mm Hg) at least 6 h apart, proteinuria greater than 5 g per 24 h, and other signs of end-organ injury*
- *Pre-eclampsia occurs in 2–8% of all pregnancies*
- *Eclampsia is defined as pre-eclampsia and a grand mal seizure in absence of other conditions account for the seizure*
- *other conditions that overlap with eclampsia:*
  - *acute ischaemic stroke*
  - *intracerebral and subarachnoid haemorrhage*
  - *cerebral venous sinus thrombosis*

- Reversible cerebral vasoconstriction syndrome (RCVS; postpartum angiopathy)develop during puerperium in the absence of hypertension or features of pre eclampsia

- Pre-eclampsia, eclampsia, and RCVS can be complicated by posterior reversible encephalopathy syndrome (PRES).

- PRES not primary diagnosis, but a clinical and imaging syndrome caused by vascular abnormalities that are present in preeclampsia and eclampsia, RCVS.

Headache

- Primary headache disorders, tension-type and migraine, most common causes of headache in pregnant and post-partum women
- Careful attention to the so-called red flags suggestive of a secondary cause
- 40% post-partum women have headaches, within the first week after delivery
- When pregnant patients get *new, worsening* headaches or when headaches *change in character*, secondary causes???????

- **Pre-eclamptic patients have bilateral throbbing headaches ass. blurred vision and scintillating scotomata.**

- **Ass. hypertension, epigastric pain, oedema, increased deep tendon reflexes, proteinuria, and agitation or restlessness**

- **Other lab findings that raise concern for pre-eclampsia include thrombocytopenia, haemoconcentration, and raised creatinine**

- ***Pregnant women with new headaches must be screened for pre-eclampsia***

- **Thunderclap headache** Abrupt onset of a severe, unusual headache need prompt investigation

- **Aneurysmal SAH** due to Hormonal changes affecting cerebral blood vessels and surges in blood pressure caused by pushing during labour

- Patients present with thunderclap headache need to exclude SAH, by CT followed by lumbar puncture if the CT is non-diagnostic.

- If work-up for SAH negative, consider
  - PRES,
  - CVT,
  - RCVS,
  - and cervicocranial arterial dissections.

- Need MRI sequences, MRA, MRV and MRI with diffusion-weighted imaging.

Post-partum primary headache study of 95 patients with severe postpartum headache,

- 37 (39%) had tension-type,
- 23 (24%) had pre-eclampsia or eclampsia,
- 15 (16%) postdural puncture headache,
- 10 (11%) had migraine,
- 3 (3%) had pituitary haemorrhage or venous sinuous thrombosis.
- 2 (1%) had mass lesions.

Postdural puncture headache

- patients given a spinal anaesthetic
- nuchal and occipital,
- caused by low intracranial pressure due to a CSF leak
- typically begin 1–7 days post partum,
- worsen upon standing
- resolve by lying flat for 10–15 mins
- symptoms usually resolve within 48 h of a blood patch procedure

Low-pressure headaches

- due to dural tears from labour-related pushing
- can cause subdural haematoma,
-
- not have the postural component of headache
- no response to a blood patch

Associated Headache Symptoms: Acute neurological deficit

- Persistent acute *motor, sensory, or visual* have more serious causes and need urgent, thorough investigations
- Pregnant with transient motor,sensory, or visual symptoms commonly have migraine with aura, even if they have no headache.
- Neurological symptoms begin gradually positive phenomena
- Positive phenomena— brightness or sparkling in vision, tingling, or prickling feelings in the limbs or body—spread gradually and lead to loss of function, such as scotoma or numbness develop and usually disappear in 20–30 min

- **Visual symptoms** are common with pre-eclampsia consider also PRES, pituitary apoplexy, and strokes.

- **Orbital haemorrhage**, diplopia, proptosis, and eye pain, and can arise during the first trimester (from hyperemesis) and during labour

- ***Stroke*** risk in pregnant and post-partum increased compared with non-pregnant especially in late pregnancy and early puerperium

  - **Rate of strokes during pregnancy and postpartum is increasing, for ICH and CVT**

- Pre-eclampsia and eclampsia have causal roles in 25–50% of patients with stroke
- Other stroke risk factors older age, hypertension and heart disease, caesarean delivery, migraine, thrombophilia, SLE, sickle cell disease, and thrombocytopenia
- HELLP syndrome (haemolysis, elevated liver enzymes, low platelets) and thrombotic thrombocytopenic purpura, increased in pregnancy and present with stroke-like symptoms

- **Cervicocranial arterial dissection** unusual cause of stroke in pregnant and post-partum women .

- present with isolated headache without neurological deficit, also have brain infarctions during the post-partum period

- **ICH and SAH**, underlying structural lesions vascular malformations and aneurysms are common

- **Vasculopathies** thrombotic thrombocytopenic purpura, pituitary apoplexy, amniotic fluid embolism, and cardioembolism from post-partum cardiomyopathy are rare causes of stroke

- Extensive diagnostic testing including vascular imaging must be done in these patients to identify specific treatable causes.

Seizures

1. most common, patients with an established seizure disorder before pregnancy

2. patients with a new non pregnancy- related seizure disorder, a new seizure from an undiagnosed brain tumour or hypoglycaemia

3. patients with new seizures that are pregnancy related (caused by eclampsia, ICH, CVT, RCVS, PRES, or thrombotic thrombo cytopenic purpura).

with PRES, seizures at presentation in absence of prodromal symptoms,

in CVT seizures usually occur later and always after headache; brain CT can be normal in both conditions

Individual conditions that cause acute neurological symptoms

Cerebral venous sinus thrombosis

- More than **75%** of cases of CVT are post partum
- Risk factors c section, dehydration, traumatic delivery, anaemia, raised homocysteine concentrations, and low CSF pressure due to dural puncture
- present with a progressively severe, diffuse, constant headache, 10% have thunderclap headache
- dizziness, nausea, seizures, papilloedema, lateralising signs, lethargy, and coma

- CT scans are often negative, but 30% of cases might show a clot or signs of infarction
- Ischaemic infarcts often undergo haemorrhagic transformation
- magnetic resonance venography is diagnostic the imaging study of choice

Reversible cerebral vasoconstriction syndrome(RCVS)

- abrupt onset of thunderclap headaches and multifocal, reversible cerebral vasoconstriction.
- Two-thirds of patients develop symptoms within 1 week of delivery and after a normal pregnancy
- associated with use of immunosuppressive drugs, serotonin reuptake inhibitors, cocaine, various other medications, blood products, craniocervical arterial dissections

- Recurring daily thunderclap headaches over several weeks after a single thunderclap headache are nearly pathognomonic
- Accompanied vomiting, confusion, photophobia, and blurred vision.
- seizures or focal neurological deficits always follow the headache.
- most patients have good outcomes, and fatal outcomes have been reported
- complications include brain haemorrhage and infarction and SAH

- Angiography after the third day usually reveals multifocal segmental arterial constriction and arterial dissections
- Non-invasive CT or magnetic resonance angiography is positive in 80% of patients,
- shows diagnostic pattern of alternating dilatation and constriction, which resembles a string of beads
- Transcranial doppler can beused to follow resolution of the vasoconstriction

Posterior reversible encephalopathy syndrome

- headache, seizures, encephalopathy and visual disturbances in setting of reversible vasogenic oedema on CT or MRI
- PRES arises in patients with acute hypertension, pre-eclampsia or eclampsia, renal disease, sepsis.
- Symptoms develop without prodrome and progress rapidly over 12–48 h.
- 90% of patients have seizures, focal to start with and then tonic-clonic,
- Severe symptoms can occur even in the absence of severe hypertension

- vasogenic oedema involves the occipital lobe,
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- 40% of patients have visual symptoms visual hallucinations, blurred vision, scotomata, and diplopia
- 1–15% of patients have transient cortical blindness
- resolve completely in hours to days; resolution of the oedema on imaging lags behind.
- EEG monitoring can detect ongoing seizure activity.
- CT will show oedema in 50–60% of patients.
- MRI reveals focal oedema, parietooccipital lobes Unlike posterior cerebral artery lesions, occipital lesions spare the medial occipital lobe and calcarine cortex
- *eclamptic patients with PRES more commonly presented with headache, less likely to be confused, and more likely to have visual symptoms.*

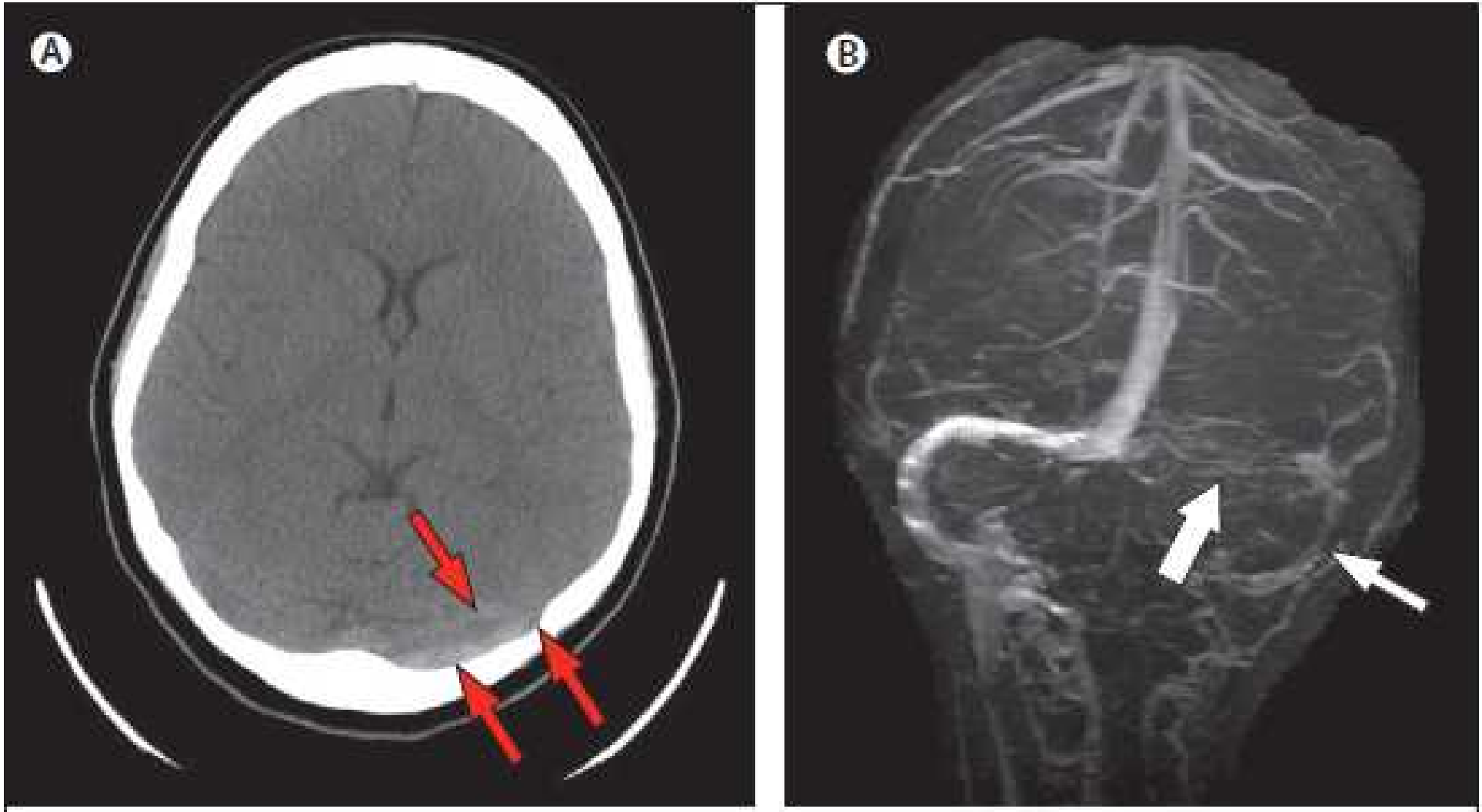
Neurological complications of eclampsia

Seizures are the hallmark of eclampsia

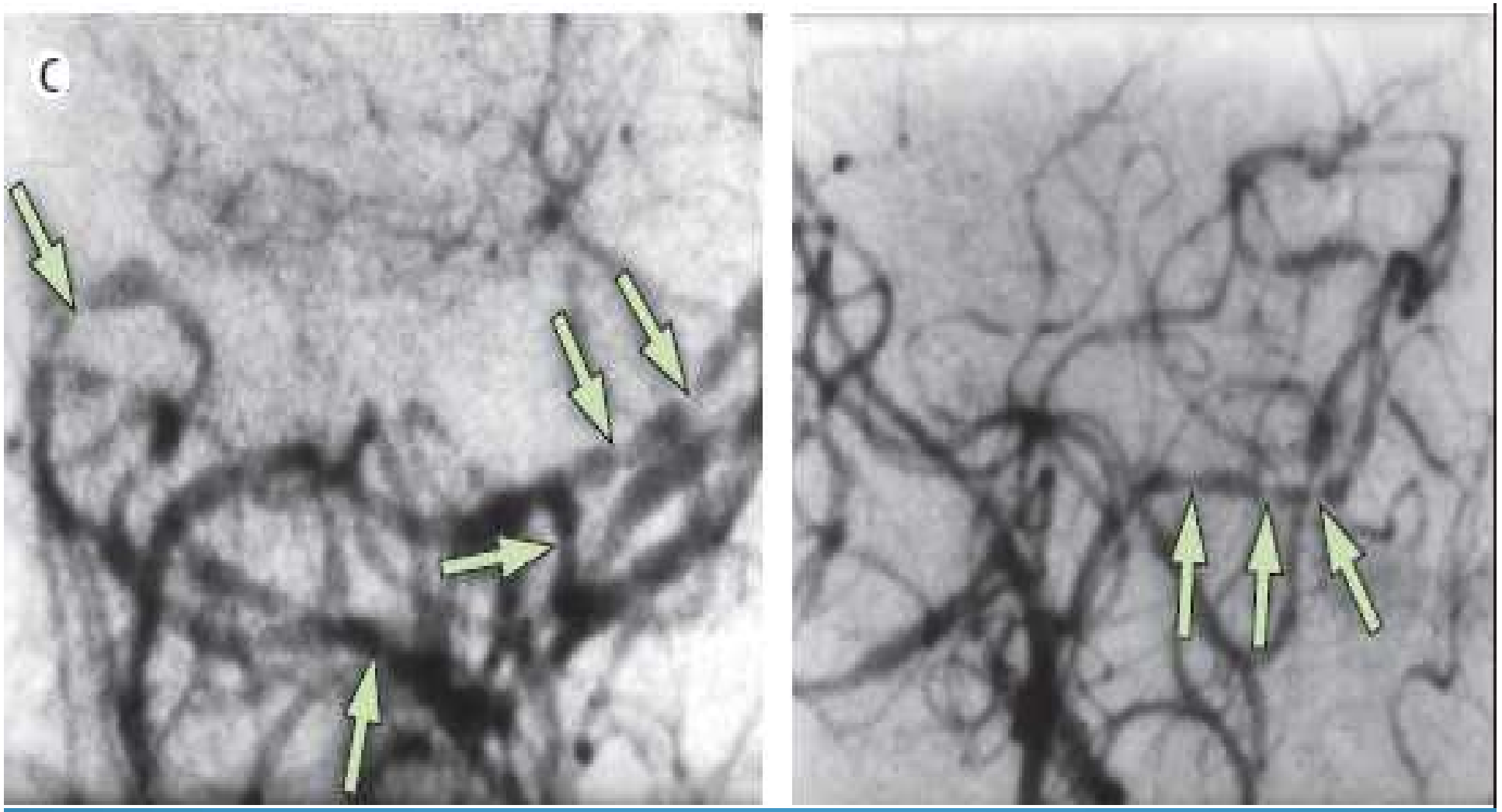
- usually tonic-clonic and last for about 1 min.
- persistent frontal or occipital headache precede seizures, blurred vision, photophobia, and altered mental status.
- In a third of cases, blood pressure is less than 140/90 mm Hg before the seizure or no proteinuria exists

- Cerebral over-regulation in response to hypertension lead to cerebral arterial vasospasm and ischaemia, result in cytotoxic oedema.
- This vasculopathy can also result in PRES or regions of infarction and haemorrhage.
- focal vasogenic oedema is characteristic of eclampsia
- Components of PRES, areas of ischaemia or haemorrhage, and even RCVS can contribute to eclamptic seizures

- over third of eclamptic seizures occur at term, and develop intrapartum within 48 h of delivery late post-partum eclampsia
- have a higher incidence of CVT, ICH, and AIS
- Range of neurological imaging findings includes infarction, haemorrhage, vasoconstriction, dissection, and both vasogenic and cytotoxic oedema.

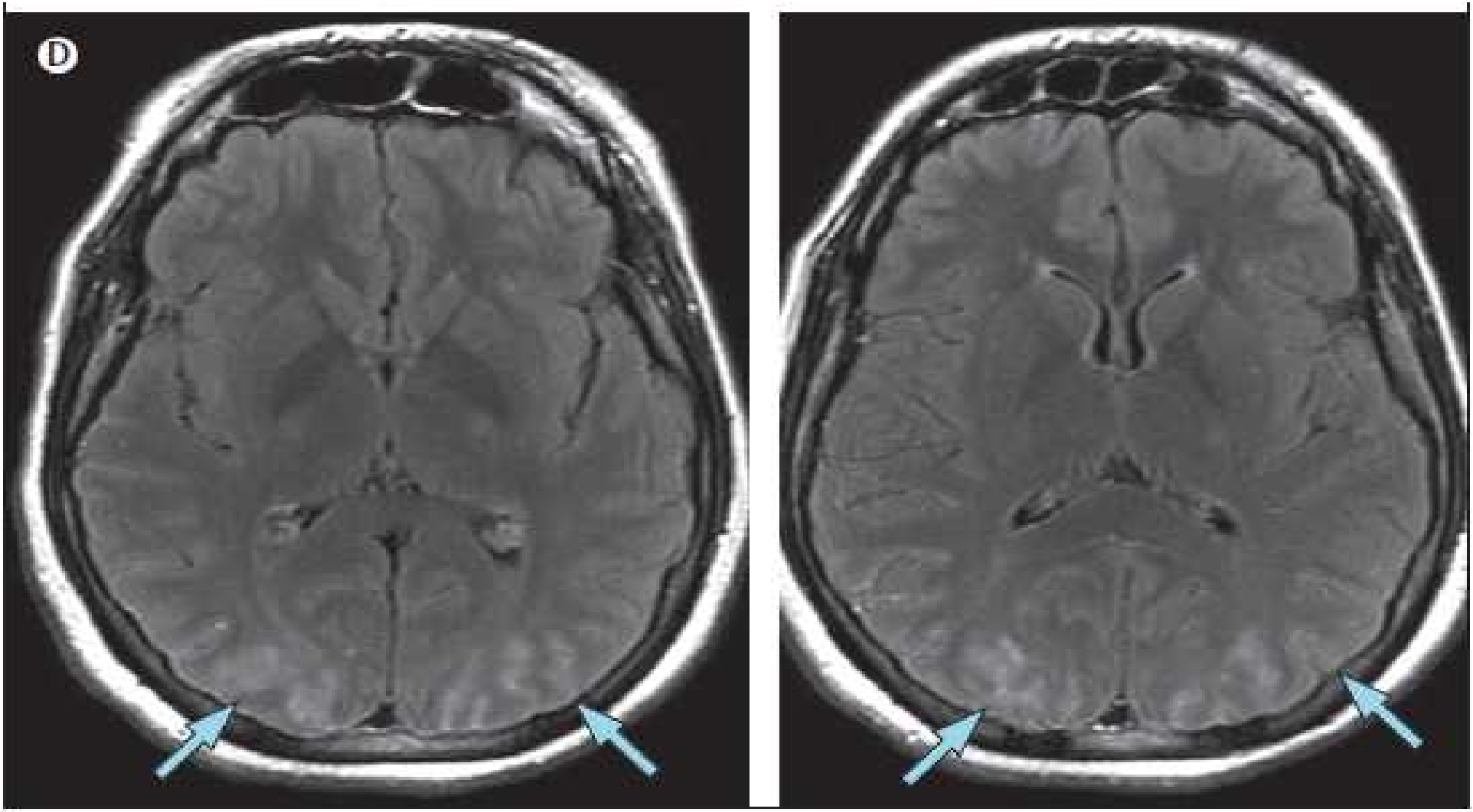


(A) Non-contrast CT scan of a 21-year-old post-partum woman who presented with 7 days of increasing left-sided headache. A subtle increased density (red arrows) that is consistent with a clot in the left transverse sinus can be seen.
(B) The magnetic resonance venogram from the same patient shows a clot in the left transverse sinus (wide arrow) and in the sigmoid sinus (thin arrow).



•Selected images from a digital subtraction angiography of a patient with reversible cerebral vasoconstriction syndrome who presented with a thunderclap headache.
•The image on the left shows the diff se nature of the vasoconstriction.

• The image on the right shows the classic so-called ***string of beads appearance*** *(green arrows show the focal areas of vasoconstriction).*



(D) Two cuts from FLAIR sequences on an MRI that show increased signal in both parieto-occipital regions (blue arrows), slightly more on the right hemisphere, in a 29-year-old pregnant patient with posterior reversible encephalopathy syndrome. Note that the findings spare the medial occipital lobe and calcarine cortex, unlike the posterior cerebral artery lesions that involve this region.

Rare conditions that cause acute neurological symptoms

- ***Amniotic fluid embolism*** causes agitation, confusion, seizures, and encephalopathy cardiovascular and respiratory collapse during or immediately after labour.
- ***Choriocarcinoma***, a rare cancer of trophoblastic tissue, metastasises to the brain cause mass effect, bleed, and invade cerebral vessels.
- ***Air embolism*** occurs when air that enters the myometrium during delivery enters the venous circulation reducing cardiac output and resulting in seizures and abnormal cognition during or just after delivery.
- air in the retinal veins and a so-called mill-wheel cardiac murmur suggest the diagnosis

- ***Wernicke's encephalopathy*** **can complicate hyperemesis gravidarum**
- **Abnormal eye movements are nearly always present**
- **the classic triad of confusion, ocular fi ndings (eg, diplopia and nystagmus), and gait abnormalities occurs**
- **the simplest test is the response to intravenous thiamine**
- ***thrombotic thrombo cytopenic purpura,*** **risk most commonly presents late in the second or early third trimester**
- **classic pentad includes thrombo cytopenia, microangiopathic haemolytic anaemia, fever, and neurological and renal dysfunction**

- fluctuating headache, seizures, and generalised and focal neurological deficits. Coexistent PRES is common
- Distinction between thrombotic thrombo cytopenic purpura (ie, plasma exchange) and HELLP (ie, magnesium and delivery of the fetus) is important.
- *Pituitary apoplexy,* acute infarction, haemorrhage of gland in(previously undiagnosed) adenoma present with headache, visual loss, and ophthalmoplegia and decreased consciousness.
- distinguished from Sheehan's syndrome (hypopituitarism presenting indolently, weeks to months after severe post-partum haemorrhage)
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- ***Chorea gravidarum*** **irregular, brief, unpredictable jerky movements of multiple body parts.**

- **associated with rheumatic fever, antiphospholipidsyndrome, stroke, Wilson's disease, and thyrotoxicosis,**

- **typically begins after first trimester, but can present post partum**

- **Symptoms usually resolve spontaneously within several weeks to months, or might subside post partum.**

Neuroimaging

Basic principles should be kept in mind

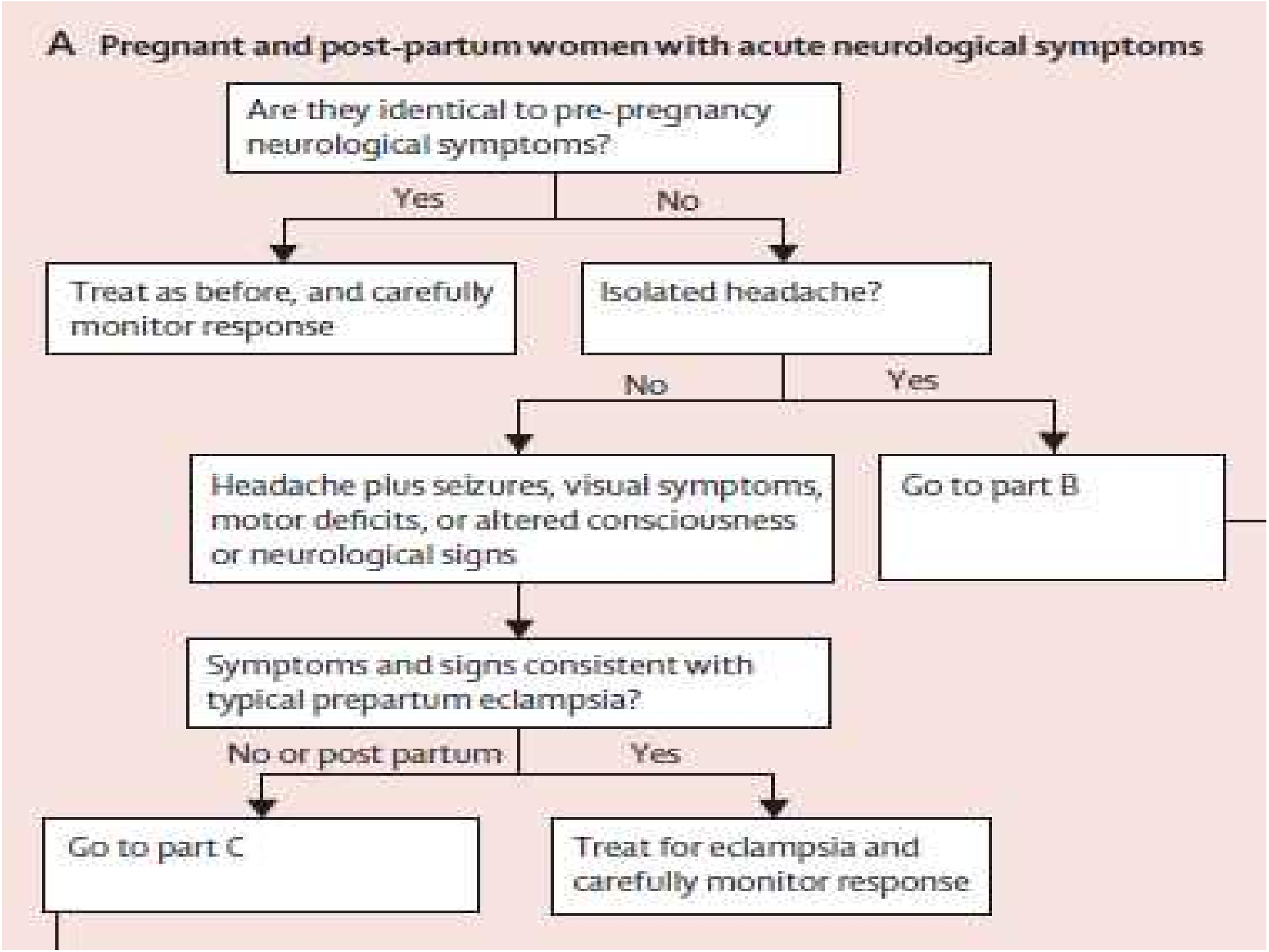
- ***clinician and the radiologist should discuss the case and differential diagnosis before imaging to minimize ionizing radiation and intravenous contrast exposure,***
- ***Second, the fetal radiation exposure from a non-contrast brain CT is negligible.***
- ***Third,MRI in pregnant patients is generally thought to be safe, although conclusive data do not exist***
- ***Fourth, as with any medication, intravenous contrast is, when possible, best avoided during pregnancy The US Food and Drug Administration classifies iodinated contrast as class B and gadolinium as class C.***

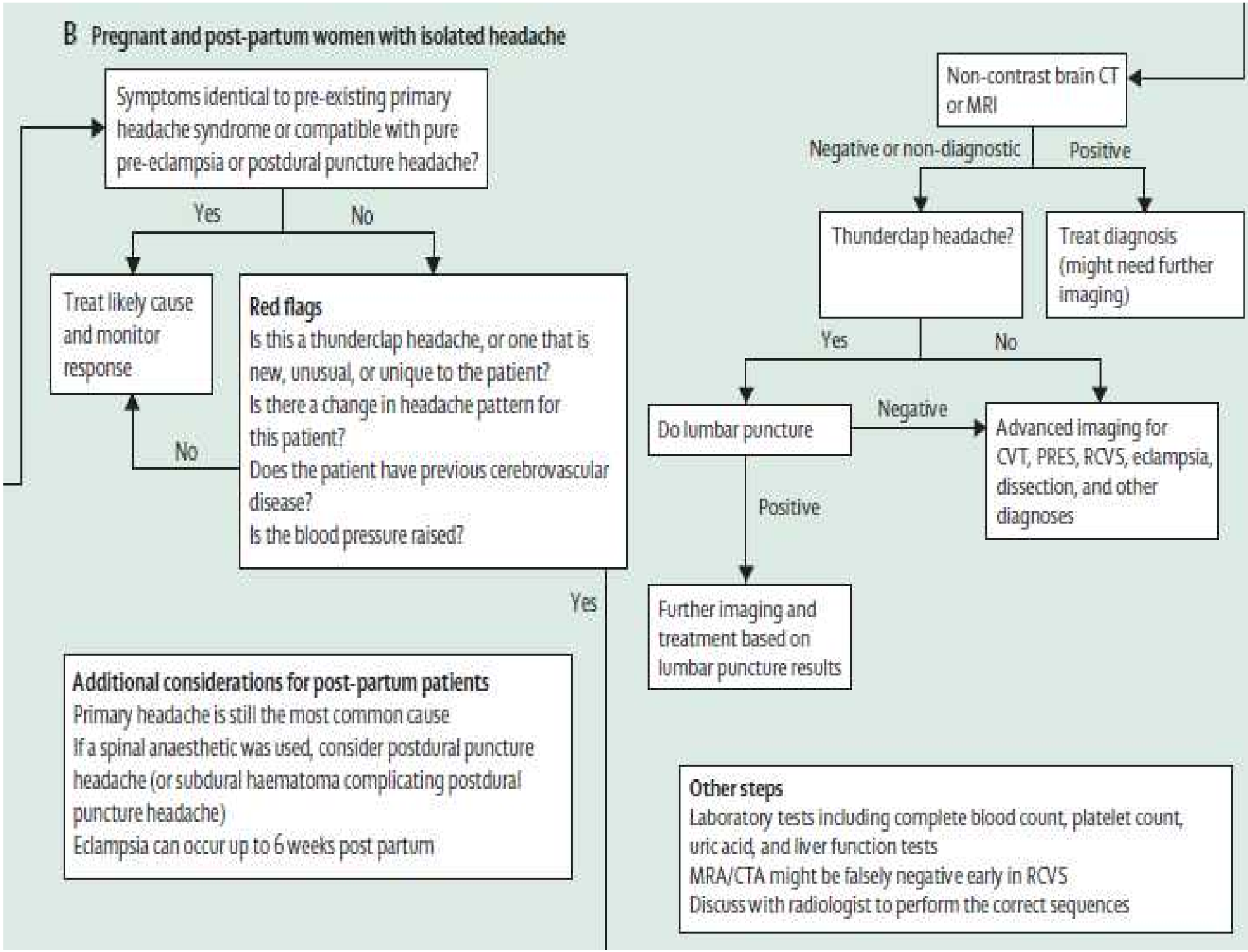
- **Recommend that *informed patient consent is obtained* before these procedures are done and intravenous contrast agents are used**

- **both types of contrast are regarded as safe in post-partum women who are breastfeeding**

Conclusion

- ***Pregnant and post-partum patients who present with new acute neurological symptoms need a thorough diagnostic evaluation that targets a range of pathological conditions that are either unique to or arise more frequently in this population.***


A Pregnant and post-partum women with acute neurological symptoms
Are they identical to pre-pregnancy neurological symptoms?
Yes
No
Treat as before, and carefully monitor response
Isolated headache?
No
Yes
Headache plus seizures, visual symptoms, motor deficits, or altered consciousness or neurological signs
Go to part B
Symptoms and signs consistent with typical prepartum eclampsia?
No or post partum
Yes
Go to part C
Treat for eclampsia and carefully monitor response


B Pregnant and post-partum women with isolated headache
Symptoms identical to pre-existing primary headache syndrome or compatible with pure pre-eclampsia or postdural puncture headache?
Yes
No
Treat likely cause and monitor response
No
Red flags
Is this a thunderclap headache, or one that is new, unusual, or unique to the patient?
Is there a change in headache pattern for this patient?
Does the patient have previous cerebrovascular disease?
Is the blood pressure raised?
Yes
Non-contrast brain CT or MRI
Negative or non-diagnostic
Positive
Thunderclap headache?
Treat diagnosis (might need further imaging)
Yes
No
Do lumbar puncture
Negative
Advanced imaging for CVT, PRES, RCVS, eclampsia, dissection, and other diagnoses
Positive
Further imaging and treatment based on lumbar puncture results
Additional considerations for post-partum patients
Primary headache is still the most common cause
If a spinal anaesthetic was used, consider postdural puncture headache (or subdural haematoma complicating postdural puncture headache)
Eclampsia can occur up to 6 weeks post partum
Other steps
Laboratory tests including complete blood count, platelet count, uric acid, and liver function tests
MRA/CTA might be falsely negative early in RCVS
Discuss with radiologist to perform the correct sequences

C Patients with other neurological symptoms or signs (with or without headache and not thought to be pure eclampsia), or eclamptic patients not responding to treatment

Appropriate consultations
Neurology and obstetrics
In some cases: critical care, neurosurgery, haematology, or endocrinology
Consider transfer to specialised centre

Advanced neuroimaging
Most of these patients will need both brain and cerebrovascular imaging by MRI

Differential diagnosis
Eclampsia
CVT
Stroke (infarct or haemorrhage)
SAH
RCVS
PRES
Subdural haematoma

Rare conditions
Choriocarcinoma
Amniotic fluid and air embolism
Pituitary apoplexy
Thrombotic thrombocytopenic purpura
Wernicke's encephalopathy


CONFERENCE ROOM
THANK YOU