Case Report

Update in
Anaesthesia

# Posterior Reversible Encephalopathy Syndrome (PRES)

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## DEFINITION

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinico-radiological syndrome of heterogeneous etiologies that are grouped together because of similar findings on neuro-imaging studies.

It was first described by Hinchey et al. in 1996 based on 15 cases[1]

This condition has been known by various names previously (reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome and reversible occipital parietal encephalopathy). PRES is now the widely accepted term.[2]

This clinical syndrome is increasingly recognized commonly because of improvement and availability of brain imaging.

### Pathophysiology

There are two main hypotheses; one involves impaired cerebral autoregulation responsible for an increase in cerebral blood flow (CBF), whereas the other involves endothelial dysfunction with cerebral hypoperfusion. Under both hypotheses, the result of the cerebral blood perfusion abnormalities is blood brain barrier dysfunction with cerebral vasogenic edema.[2]

### Clinical findings

It presents with rapid onset of symptoms including headache, seizures, altered consciousness, and visual disturbance. It is often associated with acute hypertension.

PRES is most commonly occuring with preeclampsia, eclampsia, hypertensive crisis, sepsis, autoimmune disease, cytotoxic chemotherapy, and transplantation.[3]

### Radiological findings

PRES has become synonymous with a unique pattern of brain vasogenic edema seen on CT or MR brain imaging studies. It was believed to consistently produce bilateral and symmetric regions of edema typically located in the white matter and predominating in the posterior parietal and occipital lobes[4], hence the name. Recently, it was found to present in a variety of radiological patterns;[2]

1. Holohemispheric watershed pattern; showing bilateral vasogenic edema in a linear pattern involving the white matter of the cerebellum, brain stem, occipital, parietal, frontal and temporal lobes.
2. Superior frontal sulcus pattern; showing bilateral vasogenic edema in a non-confluent patterns involving the frontal sulcus area and, to a lesser degree, the white matter of the parietal, occipital, and temporal lobes.
3. Dominant parietal-occipital pattern 'the classic pattern'; showing bilateral vasogenic edema in the white matter of the occipital and parietal lobes.
4. Partial and/or asymmetric expression of the three primary patterns.

### Differential Diagnosis

The non-specific clinical manifestations and multiplicity of radiological patterns raise diagnostic challenges. Many conditions may resemble PRES including;[2]

1. Ictal or post-ictal state (with or without status epilepticus),
2. Progressive multifocal leukoencephalopathy (PML),
3. Severe leukoaraiosis,
4. Cerebral autosomal dominant arteriopathy with subcortical infarcts,
5. Infectious encephalitis,
6. Acute disseminated encephalomyelitis,

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7. Mitochondrial myopathy encephalopathy lactacidosis and stroke-like episodes syndrome (MELAS),
8. Vasculitis,
9. Creutzfeld-Jakob disease,
10. Cerebral venous sinus thrombosis,
11. Ischemic stroke (watershed or posterior cerebral artery territory).

**Prognosis**

If promptly recognized and treated, the clinical syndrome usually resolves within a week, and the changes seen in magnetic resonance imaging (MRI) resolve over days to weeks. Patient usually recovers but permanent disability is possible.[5]

**Case Report**

A 34-year-old female presented to the emergency department 5 days after uneventful normal delivery complaining of neck pain and headache. The patient did not receive epidural analgesia for her delivery. She has no medical or surgical history of significance.

Blood investigations revealed raised WCC (12.7/uL), CRP (51.9mg/L) and D-dimer (19.8ug/ml). She had been discharged home after 2 days as all investigations including CSF analysis, CTPA and MRI brain appeared to be normal.

3 days later, she was brought again to emergency department with 2 episodes of tonic clonic convulsions and hypertension with protein in urine of +1. MRI was done showing multifocal vasogenic edema in the subcortical white matter consistent with posterior reversible encephalopathy syndrome (PRES).

The patient symptoms had improved over 3 days of receiving treatment in the intensive care unit with anticonvulsants and antihypertensives. She was then transferred to the normal ward where she repeated MRI brain which revealed significant improvement of radiological picture.

**Management**

Management of the syndrome is focused on the treatment of the underlying condition (e.g. eclampsia) and supportive treatment such as anticonvulsive agents.

**Discussion**

Posterior reversible encephalopathy syndrome (PRES) diagnosis basically depends on specific radiological features and clinical findings. The exact pathophysiological mechanism of the syndrome is still not fully understood but it may be explained by disruption of endothelial blood - brain barrier which leads to fluid and protein transudation in the brain.

High level of suspicion and early management are the key elements of recovery.

Although our case did not suffer of pregnancy related hypertension or preeclampsia during her pregnancy, hypertension was discovered on the 10th day post-delivery after her second presentation to A&E. This raises the concern of following up parturients post-delivery for the possible development of new onset preeclampsia. It is worth mentioning that preeclampsia can present as late as 4 to 6 weeks postpartum according to American College of Obstetricians and Gynecologists guidelines.[6]

PRES can be one of the complications that could result from untreated preeclampsia. Better outcome can be attained by early diagnosis and prompted treatment.

## CONCLUSION

PRES should be considered in patients who present with seizures, altered consciousness, visual disturbance, or headache, particularly in the context of acute hypertension. It should not be missed as one of differential diagnoses of postpartum headache and convulsions. Fortunately, if it is promptly recognized and treated, the clinical syndrome usually resolves within a week.

## REFERENCES


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