

# Challenges faced during Obstetrical Management in Pregnant Woman with Rare Neurological Conditions: A Retrospective Study

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## ABSTRACT

**Aim:** Several neurological disorders occur in women of the childbearing age group and are an important cause of maternal morbidity and mortality. The diagnosis and management of neurological disorders in pregnancy is always a challenging task due to varied symptomatology and risks to the fetus. A rare neurological disorder in pregnancy can be pregnancy related or can be caused by exacerbation of a pre-existing neurological condition or sometimes can be detected for the first time during pregnancy which might be an incidental finding. Treating these patients remain a challenge. Thus, our study aimed to determine the challenges in managing pregnant mothers with rare neurological complications in a tertiary health care hospital.

**Methods:** All pregnant women with rare neurological conditions admitted to the Department of Obstetrics & Gynaecology, St. Johns Medical College and Hospital, Bengaluru, Karnataka, India during our 1-year study period (from January 2020 to December 2020) were analyzed retrospectively. Our experience may help to take proper decisions when a similar condition is encountered elsewhere.

**Results:** In a study period of 1 year, the total incidence of neurological disorders with pregnancy was 0.89%, which included eclampsia (0.4%) and epilepsy (0.25%).

We had nine cases with a rare neurological disorder. Three of them were primigravida (33.5%). Four were diagnosed with the neurological disorder before the pregnancy. Five cases were diagnosed for the first time during pregnancy. We had two cases with Wernicke's encephalopathy (WE), one case of the acute motor sensory axonal neuropathy (AMSAN) variant of Guillain-Barré syndrome (GBS), and one case with GBS. One case with right cerebellopontine angle tumor which got diagnosed during her first trimester of pregnancy. Four cases with pre-existing neurological disorder included status epilepticus, limb-girdle muscular dystrophy, middle cerebral artery (MCA) aneurysm/internal carotid artery (ICA) supraclinoid occlusion and situs inversus totalis with neuromyelitis optica.

The mean maternal age was 27.77 years. Gestational age varied from case to case at the time of their presentation and five cases had associated obstetric complications.

Six mothers recovered well, one mother went against medical advice, two mothers lost to follow-up and we had one maternal mortality.

**Conclusion:** Even though knowledge about rare neurological diseases in pregnancy is finite, these cases require tertiary care referral and patients should be promptly managed through a multidisciplinary approach. Strict antepartum vigilance is essential for the treatment and good neonatal outcomes. Holistic and vigilant care toward these patients reduces mortality and improves maternal and neonatal outcomes.

**Keywords:** Cerebellopontine angle tumor in pregnancy, Guillain-Barré syndrome in pregnancy, Limb-girdle muscular dystrophy, Neurological disorder, Pregnancy, Wernicke's encephalopathy.

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## INTRODUCTION

Several neurological disorders occur in women of the childbearing age group and are an important cause of maternal mortality. It has been known to account for nearly 7% of maternal deaths.<sup>1</sup> Such disorders are encountered especially during pregnancy; however, a few neurological disorders may have increased frequency of presentation in pregnant women; for example, Bell's palsy, specific types of strokes, and benign intracranial hypertension or pseudotumor cerebri and epilepsy.<sup>1</sup> Some of them precede pregnancy and others have their new onset of symptoms during pregnancy and these must be distinguished from pregnancy complications.<sup>1</sup>

A rare neurological disorder in pregnancy can be pregnancy related or can be caused by exacerbation of a pre-existing neurological condition or sometimes can be detected for the first time during pregnancy in which it might be an incidental finding.<sup>2</sup> The diagnosis and management of neurological disorders in pregnancy is always a challenging task due to varied

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symptomatology and risks to the fetus.<sup>2</sup> More importantly, pregnancy can predispose to the onset or deterioration of pre-existing neurological disorders, marking a

wide index of suspicion.<sup>2</sup> Most of these conditions are infrequent and require a specialized and multidisciplinary management.

Pregnant women most often experience sensory abnormalities which resolve soon after delivery.<sup>3</sup> Treatment is challenging due to risks to the unborn child.<sup>3,4</sup> Therefore, a basic knowledge of the modifications that affect the nervous system, including neuroanatomy, reproductive endocrinology, immunological system, and the wider role of sex steroids in many of these adaptations can guide clinicians in the diagnosis and management of the pregnant patient with neurological symptoms.<sup>3,4</sup>

Guillain-Barré syndrome is an immune-mediated peripheral neuropathy characterized by rapidly evolving polyradiculoneuropathy.<sup>5</sup> It is generally preceded by a triggering event, most often an infection, flu-like illness or gastroenteritis.<sup>5</sup> Its estimated incidence in pregnancy 1.2–1.9%.<sup>5</sup> The management in pregnancy is similar to that in the non-pregnant population which includes intravenous immunoglobulins (IVIG), plasmapheresis, and ventilator support if required.<sup>5</sup> Studies show that the risk of GBS increases in the puerperium and reports of relapse have been documented.<sup>5</sup> Hence, early diagnosis, a multidisciplinary approach, and prompt immunomodulatory therapy are the cornerstones in the management of GBS during pregnancy to improve outcomes for the mother and fetus.<sup>5,6</sup>

Limb-girdle muscular dystrophies are genetically determined progressive skeletal muscle disorders ranging from rapid progression to milder onsets with its manifestations in first few decades or with milder forms of late onsets.<sup>7</sup> Its extremely rare disorder with its incidence being less than 1 per 1-lakh population. Genetic mutations screening for defective protein expression is an important tool.<sup>7</sup> The dilemma in choosing the mode of delivery and pregnancy outcome remains hypothetical.<sup>7</sup>

Status epilepticus is a rare presentation in pregnancy, the most common etiology of neurological disorder in pregnancy being eclampsia<sup>8–10</sup> (1 in 200 pregnancies).<sup>10</sup> The challenge in managing these patients requires prompt treatment with a multidisciplinary approach and a detailed knowledge about antiepileptics and anesthetic agents.<sup>8–10</sup>

Wernicke's encephalopathy is a reversible yet serious neurological manifestation caused by vitamin B1 (thiamine).<sup>11</sup> In pregnancy, it is commonly associated with hyperemesis gravidarum (HG), starvation, and prolonged intravenous feeding. Its serious complications being central pontine myelinolysis (CPM) due to electrolyte fluctuations (hypokalemia, hypernatremia, and hyperosmolality). Continuing the pregnancy with this condition is a challenge, and needs a multidisciplinary approach.<sup>11</sup>

The incidence of cerebellopontine (CP) angle tumor in pregnancy is very rare. Incidence of brain tumor in pregnancy is 6 in 1-lakh pregnancies, thus its aims at significant challenge to diagnosis and manage these patients during pregnancy.<sup>12</sup> Treatment options depend on the patient's stage of pregnancy and neurologic status and the treatment strategy involving a multidisciplinary team of obstetrics, anesthesia, and neurosurgery is ideal.<sup>12</sup> Resection of tumor in pregnancy is mandatory and the optimal time for surgery is during second trimester.<sup>12</sup>

Incidental aneurysm and its effects such as bleeding, rupture, and increase in size remains uncertain. Aneurysmal subarachnoid hemorrhage accounts for up to 1 in 10 of all maternal deaths in pregnant women and 1 in 4 maternal deaths from indirect causes. The challenge is in planning normal delivery depends on the corrective surgeries done previously. A cesarean delivery is recommended out of concern about the effect of increase

in intracranial pressure during delivery; however, there are no direct evidence available. The maternal mortalities after cesarean delivery are very low.<sup>13</sup>

Thus, our study was aimed to determine the challenges in managing the pregnant mothers with a rare neurological complication in a tertiary health care hospital.

## MATERIALS AND METHODS

In this retrospective case analysis, all pregnant women with rare neurological conditions admitted to the Department of Obstetrics & Gynaecology, St. Johns Medical College and Hospital, Bengaluru, Karnataka, India during our 1-year study period (from January 2020 to December 2020) were analyzed.

## RESULTS

In a study period of 1 year, the total incidence of neurological disorders with pregnancy was 0.89%, which included eclampsia (0.4%) and epilepsy (0.25%).

We had nine cases with rare neurological disorder. Three of them were primigravida (33.5%). Four were diagnosed with neurological disorder before the pregnancy. Five cases were diagnosed for the first-time during pregnancy (Table 1).

We had two cases with WE diagnosed with the help of magnetic resonance imaging (MRI) brain, one case of AMSAN variant of GBS, and one case with GBS. One case with right cerebellopontine angle tumor which got diagnosed during her first trimester of pregnancy (Table 1).

Four cases with pre-existing neurological disorder included status epilepticus, limb-girdle muscular dystrophy, MCA aneurysm/ICA supraclinoid occlusion, and situs inversus totalis with neuromyelitis optica (Table 1).

**Table 1:** Demographic data

Age of mother (mean ± SD)	27.77 (5.562)
Pre-existing neurological disorder	4 (44.5%)
Neurological disorder diagnosed during pregnancy for the first time	5 (55.5%)
Associated obstetric complications	5 (55.5%)
Mode of delivery	
Vaginal	2 (22.2%)
LSCS	4 (44.4%)
(three cases had loss of follow-up)	

**Table 2:** Maternal outcome

Recovered	6 (66.6%)
Mortality	1 (11.1%)
Lost to follow-up	2 (22.2%)

**Table 3:** Neonatal outcome

Preterm	3 (33.3%)
Term	3 (33.3%)
NICU admission	3 (33.3%)
Fetal demise	1 (11.1%)

**Table 4:** Description of case population with neurologic disorders

S.No.	Age (years)	Gestational age (weeks)	Obstetric score	Diagnosis	Obstetric complications	Medical complications	Treatment	Mode of delivery	Maternal outcome	Fetal outcome
1.	29	27	Primigravida	Status epilepticus	Nil	MRI showing leukodystrophy, MRI spectroscopy showing glioma	Antiepileptics		DAMA	Not known
2.	20	29 + 2	Primigravida	AMSAN variant of GBS	Preterm	Hyperthyroidism	IVIG, carbimazole	Vaginal	Restoring activity of daily living by physiotherapy	Preterm care NICU
3.	26	28	G3P2L2	GBS	Nil	Aspiration pneumonia	IVIG	Lost to follow-up	Recovered	Lost to follow-up
4.	33	38 + 2	Primigravida	Limb-girdle muscular dystrophy	Non-severe PE	Nil	Carnitol	Elective LSCS under GA	Restored daily activities	Healthy baby
5.	30	16	G3P2L1D1	WE	Intrauterine fetal demise	Encephalitis	Injection thiamine	Vaginal	Recovered	Fetal demise
6.	21	17 + 3	G2A1	WE	Nil	Severe anemia	Treated outside with injection thiamine	Lost to follow-up in view of COVID-19	Not known	Not known
7.	24	28	G2P1L1	Right cerebellopontine angle tumor	Nil	Aspiration embolism/pneumonitis	S/P VP shunting	Emergency LSCS	Maternal death	Preterm care NICU
8.	30	34 + 1	G3P1L1	MCA aneurysm/ICA supra clinoid occlusion	Previous LSCS/reduced AFI 8 cm	Nil	Injection levipill	Elective LSCS	Recovered	Healthy baby
9.	31	37	G3P1L1A1	Situs inversus totalis with neuromyelitis optica	Increased AFI 22 cm, macrosomia gestational diabetes melitus	Nil	Tablet wysolone Tablet azoran Tablet carbamazepin Tablet baclofen Tablet gabapentin	Elective LSCS	Restored daily activities	Baby congenital hypothyroidism

AFI, amniotic fluid index; DAMA, discharge against medical advice

The mean maternal age was 27.77 years; gestational age varied from case to case at the time of their presentation and five cases had associated obstetric complications (Table 1).

Six mothers recovered well by the time of discharge. We had one maternal mortality (case of right CP angle tumor), one mother got discharged against medical advice (case of status epilepticus) and two mothers lost to follow-up in view of the COVID-19 lockdown (Table 2).

Three of the neonates required neonatal intensive care (NICU) mainly for the preterm care. One mother with WE had fetal demise (Table 3).

Complete information about all the cases have been listed in Table 4.

## DISCUSSION

Although neurological illness in pregnancy has varied symptoms, it warrants an urgent neurological review.<sup>2</sup> A medical dilemma exist to this very day to make an adequate diagnosis in the management of such rare illness.<sup>2</sup> A detailed history and sophisticated investigation (MRI) aid in diagnosis of such high-risk pregnancies.<sup>2</sup> Incidence of neurological diseases in pregnancy happen to be highly varied due to the influence of the hormones, and its exaggerated physiological responses that mediate various enzyme process.<sup>2</sup> Hence, a multidisciplinary approach is mandated throughout the course of pregnancy and suggests immediate referral to a high-risk maternity center, with a good NICU back-up.<sup>4</sup>

Our study highlights the challenges faced during the management of neurological disorders in young females with pregnancy in the age groups of 20–37 years.<sup>2</sup> A study done by Renukesh and Rai predicted the manifestation of neurological diseases seem to occur in primigravida, most of them without pre-existing neurological symptoms.<sup>2</sup> However, in our study, 6 cases were of multigravida, 3 were of primigravida, and 55.5% of them were of without pre-existing neurological disorder.

All patients were referred to our tertiary center, and underwent MRI modality of testing. The termination of pregnancy was determined by consideration of the individual cases. Elective lower segment cesarian section (LSCS) under general anesthesia was conducted in one case of limb–girdle muscular dystrophy as per neurologist opinion and was found to have good pregnancy outcome.<sup>6–8</sup>

Literature search for the mode of delivery in such cases did not give clear guidelines or conclusions. There is one study reporting successful vaginal delivery.<sup>14</sup> Other studies concluded that management of limb–girdle muscular dystrophy should be individualized, as the symptoms vary among patients and spinal anesthesia can be safely used. However, in our study, a case of limb–girdle muscular dystrophy was managed by elective LSCS as per neurologist advice under general anesthesia there were no complications observed during LSCS or in the postoperative period. Mother did not develop progressive weakness during recovery phase.

Majority of the patients required an intensive care unit admission and prolonged hospital stay with a good medical management with antiepileptic drugs and neuroprotective agents like carnitine.<sup>6</sup> We had 1 case with glioma which presented in antenatal period during the evaluation of antepartum status epilepticus at gestational age of 27 weeks.<sup>6</sup> This clinical entity has a low incidence in certain population as seen with a French, Brazilian, and UK study.<sup>6</sup>

As pregnant women have a physiological transitory state of immunotolerance which prevents the rejection of the fetus, there is a remit of Type 1 T helper (TH1)-cell-mediated autoimmune disease.<sup>9</sup> A varied phenomenon of this physiological process takes place in GBS which requires IVIG.<sup>9</sup> Our study showed an adequate good clinical response to IVIG, suggesting the benefits of such treatment modalities in pregnancy.

Despite adequate and timely counselling regarding the morbidity associated maternal and neonatal outcome, patient went against medical advice after receiving a good initial evaluation and treatment and two cases were lost to follow-up.<sup>15</sup> This mandates a need for new government schemes incorporating the inclusion of neurological health and subsidized treatment beneficiaries, which can be incorporated into maternal and child health (MCH) programs in the future.

## CONCLUSION

Neurological diseases in pregnancy are complex and need utmost clinical vigilance. The MRI tests still seems to be the gold standard modality of confirming diagnosis. Choice for the mode of delivery is restricted to the clinician perspective of the individual case at the time of presentation. A strict antepartum vigilance is essential for the management and good neonatal outcome. Family counselling of such neurological cases at the time of admission plays a vital role in prevention of loss of cases to follow-up.

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