



Case Report

Management considerations in Addison's disease complicating pregnancy

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ABSTRACT

Addison's disease is a rare endocrine disorder. We present the management of a pregnant lady with Addison's disease. She was managed jointly with the endocrinologists and the titrated dose of hydrocortisone and fludrocortisone dosage was regulated. Her antenatal care was uneventful and she had emergency caesarean section at 39 weeks in view of abnormal CTG. Her labour was appropriately covered with rescue steroids. However she went into hypotension immediately after delivery. She was resuscitated and was managed further in ICU where high dose steroids were given. She made an uneventful recovery.

Key Messages: 1. Management of pregnancy with Addison's disease is challenging but are often achieved with appropriate multidisciplinary management; 2. Patients with Addison's disease tolerate pregnancy well if the replacement steroids are adjusted and monitored closely; 3. Labour and delivery need to be managed cautiously with adequate steroid replacement to ensure successful outcomes; 4. Differentials need to be considered when adequate steroid cover has been provided and still there is a picture of Addisonian crisis.

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1. Introduction

Addison's disease (Primary adrenal insufficiency) is a rare endocrine disorder of adrenal cortex where there is insufficient production of glucocorticoids and mineralocorticoids. Addisonian crisis is a life threatening event and may accompany stressful conditions. Pregnant women with Addison's disease need to be managed with care as crisis can occur in situations like labour, puerperium, hyperemesis gravidarum. Here we discuss the management issues around pregnancy and delivery by presenting the case of a pregnant lady with Addison's disease who also developed severe hypotension post-delivery.

2. Case History

A 25year old primigravida, a known case of Addison's disease booked for her antenatal care at our hospital at 12 weeks of gestation. She was diagnosed as Addison's disease three years back during her evaluation for excessive

tiredness and hyperpigmentation. Her Serum cortisol levels at 30 and 60 minute post 250 mcg IV ACTH was 4.7 and 4.9 mcg/dl (Normal-18mcg/dl) respectively with normal electrolytes, thyroid functions, and adrenals on imaging. She was commenced on Hydrocortisone 20mg/day and Fludrocortisone 100mcg/day replacement and was stable. Her pregnancy was jointly managed with endocrinologist. She was continued on her steroid replacement dose where in Hydrocortisone was gradually increased to 40mg/day by the end of third trimester and she was monitored closely for signs and symptoms of under and over steroid replacement. She had an uneventful pregnancy with normal growth scans.

She went into spontaneous labour at 39 weeks gestation. She received adequate stress cover steroids (Inj. Hydrocortisone 100 mg 8th hourly for cover during labour). She needed emergency LSCS for CTG abnormalities. She had epidural analgesia but needed spinal anaesthesia as epidural top-up did not work. However per operatively she developed profound hypotension (unrecordable BP, bradycardia) and was resuscitated with IV fluids, vasopressors, steroids and ventilated.

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Post operatively in ICU she developed neurological deficits (disorientation, slurring of speech, ataxia) on Day 1. MRI revealed undiagnosed Arnold Chiari malformation with cerebellar tonsillar herniation (CTH). She was managed conservatively and neurological deficits were completely reversed by Day 3.

3. Discussion

The majority of women with Addison's disease have normal pregnancy outcomes, and overall risks are generally considered to be low as per the population based study from Sweden. Successful outcome in pregnant women with Addison's disease are often achieved by a multidisciplinary model of care with careful monitoring of foetal growth.¹

Pregnancy and labour in these women need to be managed with adequate steroid coverage and close monitoring. During normal pregnancy, circulating cortisol concentration is increased 2- to 3-fold, with a continuous increase from the first trimester onward due to increases in CBG levels. Hence there is a need to increase hydrocortisone dose by 20–40% from the 24th week onward to reflect the physiological cortisol levels. Hydrocortisone is generally preferred over other steroids and Dexamethasone is usually contraindicated because it crosses the placenta. Close monitoring for clinical symptoms and signs like normal weight gain, fatigue, postural hypotension or hypertension, hyperglycemia has to be made. These indicate the adequacy of the glucocorticoid dosage. Mineralocorticoid requirements during pregnancy are harder to assess but serum electrolyte levels help in the assessment. Though there may be a need to increase the fludrocortisone dose this is covered by the increase in hydrocortisone in most cases.²

Adrenal crisis can happen due to insufficient glucocorticoid dosage or due to stressful situations. Addisonian crisis which is a life threatening event can occur in pregnancy in situations like labour, puerperium, hyperemesis gravidarum. Adequate stress cover steroids need to be provided to prevent crisis.³ A bolus injection of 100 mg hydrocortisone IV followed by continuous infusion of 200 mg hydrocortisone over 24 hours has been recommended. This case emphasises the necessity for considering differential diagnosis for Addisonian crisis.

Our patient received appropriate steroid replacement in pregnancy as well as adequate steroid cover during labour. Peroperatively she developed profound hypotension and bradycardia and this could be due to Addisonian crisis but possibility of high spinal hypotension need to be considered, especially in the background of adequate maintenance and stress glucocorticoid coverage. Her neurological symptoms were possibly due to the aggravation of the tonsillar

herniation due to cerebrospinal fluid leak after the spinal.

Addison's can be due to a variety of causes though autoimmune adrenalitis and Tuberculosis in developing countries account for the majority of adult cases.³ A rare entity, Triple A (Allgrove) syndrome has been described where there is association of Addison's disease with Arnold Chiari malformation. Achalasia and alacrimia are also associated and it is due to autosomal recessive inheritance of triple A syndrome gene, designated AAAS located on chromosome 12q13.⁴

4. Source of Funding

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5. Conflict of Interest

None.

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