Addison’s Disease in Pregnancy

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I. Introduction

• Addison's disease (Primary adrenal insufficiency) is a rare and chronic disease of adrenal cortex where there is insufficient production of glucocorticoid and mineralocorticoid.

• Addisonian crisis, a rare but life-threatening event in pregnant women, may accompany stressful conditions such as labor, puerperium, infection, hyperemesis gravidarum or surgery.

• Addison's disease has deleterious effects on pregnancy outcome. It may cause infertility, abortion, intrauterine growth retardation, intrauterine fetal death and postpartum adrenal crisis.

• Adrenocortical insufficiency is related to a higher incidence of serious fetal complications like intrauterine death.

• With introduction of glucocorticoid therapy, pregnancy has become less risky for Addisonian women.

• Maternal mortality has been reduced from 45% of 1930 to 7% of 1948-1955 until to 0.7% of 2000.

• We represent a case of Addison's disease in pregnancy where pregnancy was brought to a successful conclusion without any undue complication by the multidisciplinary management.

II. Case Report

• A 24 year Primigravida @ 38w + 2days gestation age with k/c/o Addison’s disease came for safe confinement.

• She was diagnosed as a case of Addison's disease for last two years and was under consultation of an endocrinologist. Two years back she was having nausea, vomiting, fatigue, weakness and loss of weight. Hyper pigmentation was also present.

• Patient's blood tests did not show the typical change in the serum electrolytes with low sodium and raised potassium levels (Sodium-137, Potassium-3.8)

• But her morning serum cortisol level was typically very low (2.90 mcg/dl) where the normal range is 4.2 - 38.4 mcg/dl).

• During her antenatal period she was managed by obstetrician and endocrinologist.

• She was on Tab. Hydrocortisone 20mg-0-10mg.

• All three trimesters were uneventful.

• Patient was induced with Pge2 gel.

• Inj. Hydrocortisone 100 mg iv stat before induction followed by 100 mg iv tds for 24 hrs to prevent addisonian crisis.

• Patient underwent caeserean section in view of fetal distress and delivered a Girl baby 3.445 kgs, Apgar 8/10, 9/10

• At follow-up both mother and baby have no health problems and maternal hormonal therapy was changed like pregravidic schedule to oral Tab.Hydrocortisone 20mg-0-10mg.

• In this case baby was free from any congenital defect and development was normal.

III. Discussion

• Pregnancy in Addison’s disease is rare.

• These patients usually suffer from subfertility.

• Cortisone appeared to cure the subfertility, well being and life span of patients with Addison’s disease.

• Pregnancy and in particular labour and its complications were hazardous in the past.(1,2).

• Diagnosis of Addison's disease during pregnancy may be difficult and requires much awareness of the physician.(7)

• Both Addison's disease and physiological changes in normal pregnancy share some common Symptom such as fatigue, nausea, vomiting, weakness, hyper-pigmentation and hypotension.2.

• In one study on 13 cases where cortisone was given(3). One case reported maternal death among these 13 cases. Two cases reported vomiting at 8 weeks of pregnancy led to adisonion crisis and managed by cortisone (4).
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• The replacement of glucocorticoid and mineralocorticoid should be continued throughout pregnancy, delivery and lactation (5).
• Dose depends on clinical condition and serum electrolyte level.
• The increase in dose is usually required during third trimester.

Management of Addison’s disease – labour and postpartum period

During Labour
• Adequate hydration should be maintained through intravenous normal saline.
• Glucocorticoid-Hydrocortisone sodium succinate is given by intravenous route at 25mg Q6th hourly (8).
• At the time of delivery if labour is prolonged, high dose parental hydrocortisone 100mg Q6th hourly should be given. (8)
• After delivery, dose is tapered to a maintenance dose in three days (8).
• BP should be monitored Q4th hourly.

Postpartum
• Satisfactory lactation depends on adequate supply of adrenal glucocorticoids.
• An adrenal crisis is more likely to develop during the first 24 hours after delivery than any other time during the child bearing incident (1). This results from inability of the adrenals to meet the stress of labour.
• The loss of biologically active placental steroids has been suggested as an additional factor, but hasn’t yet been proved.
• The blood pressure as determined by hourly reading is the best guide to the adequacy of the dosage of cortisone, a fall indicating the need for further hydrocortisone.
• After the first 2 days of the puerperium the dose of hydrocortisone is gradually reduced to the previous maintenance level.

IV. Conclusion

• A pregnant women with Addison’s disease is a high risk pregnancy and multidisciplinary approach is needed.
• Adequate steroid replacement along with regular follow up before and during pregnancy may allow normal fetal growth and prevent complications of pregnancy, Labour and puerperium.

Reference
[7]. Sara Bird. Failure to diagnose addisons disease, professional practice: Risk management, MDA National.