International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Swamy M et al. Int J Reprod Contracept Obstet Gynecol. 2026 Jan;15(1):315-318
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20254302

Case Report

Pregnancy outcome in a woman with congenital adrenal hyperplasia: a rare case report

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Received: 16 October 2025
Revised: 28 November 2025
Accepted: 29 November 2025

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ABSTRACT

Congenital adrenal hyperplasia (CAH) refers to family of inherited disorders of adrenal steroidogenesis caused by mutations in genes encoding steroidogenic enzymes involved in glucocorticoid synthesis/in cofactor enzyme P450 oxidoreductase that serves as electron donor to CYP21A2 and CYP17A1 with autosomal recessive inheritance pattern. A 24-year-old primigravida with simple virilizing CAH who underwent reconstructive surgery in childhood. During pregnancy, she developed severe preeclampsia requiring preterm emergency caesarean delivery at 30+6 weeks period of gestation. Stress steroid supplementation was administered intrapartum. Both mother and neonate had a favourable outcome with close multidisciplinary management. Early diagnosis, timely reconstructive surgery, optimized steroid replacement, and multidisciplinary care are crucial for favourable maternal and neonatal outcomes in CAH pregnancies.

Keywords: Congenital adrenal hyperplasia, Pregnancy, Steroid replacement

INTRODUCTION

Congenital adrenal hyperplasia (CAH) comprises a group of autosomal recessive disorders. Incidence of classical CAH ranges from 1 in 10,000 to 1 in 20,000 live births with carrier rate of around 1 in 60.[1] It is caused by defects in adrenal steroidogenesis, most commonly due to 21-hydroxylase deficiency. It leads to impaired cortisol synthesis, androgen excess, and variable aldosterone deficiency.[2] Management of CAH requires a multi-disciplinary approach. Here we bring out the complexities in managing a pregnant woman with CAH.

Table 1: Chance of child being affected.[1]

Partner 1	Partner 2	Chance of Child being affected	Chance of female child being affected
Carrier	Carrier	One in four	One in eight
Affected	Carrier	One in two	One in four

CASE REPORT

Unbooked, 24-year-old Primigravida with 30+6 weeks of gestation with simple virilizing congenital adrenal hyperplasia came to the OPD for regular antenatal checkup.

She had undergone vaginoplasty and clitoroplasty in 2014 for ambiguous genitalia. Karyotype was 46 XX. She was on corticosteroid replacement with Prednisolone and Fludrocortisone up to 9 years of age, which was replaced with oral Betamethasone for maintenance.

Ultrasound revealed normal female internal organs. Hence, steroids were switched over to Prednisolone at 12 years of age. She has had regular menstrual cycles.

No significant family history.

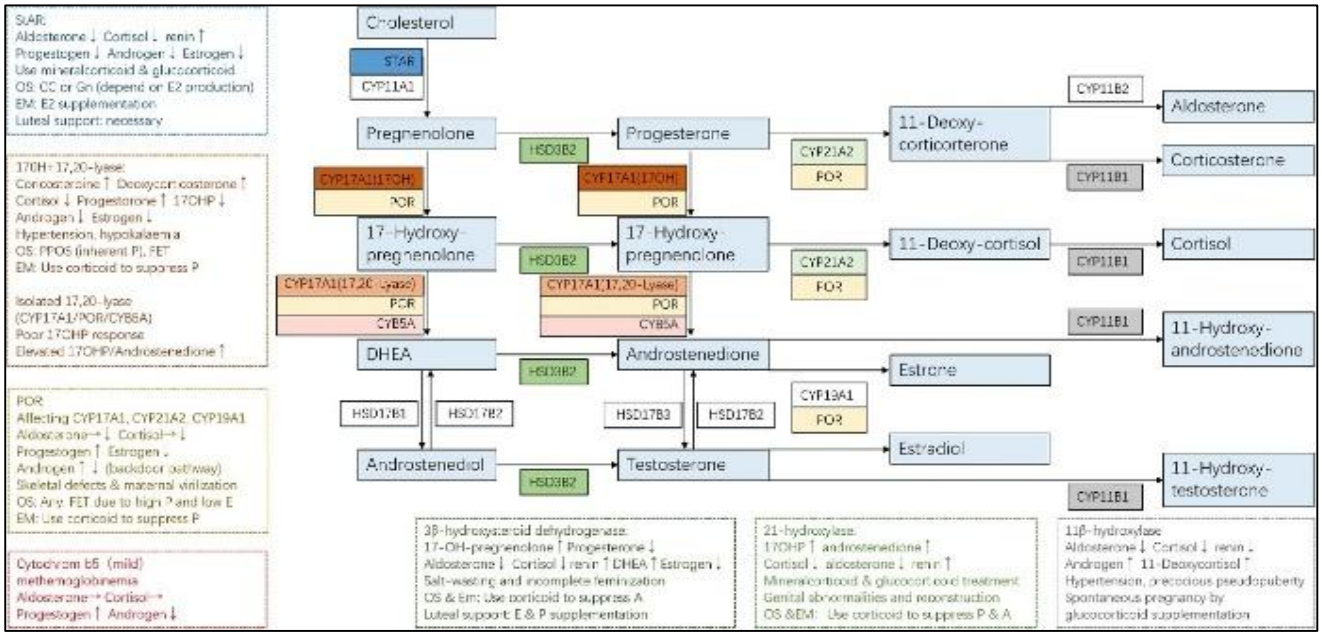

Figure 1: Adrenal steriodogenesis.[3]

She was on oral betamethasone in the preconceptional period with escalation of dose on pregnancy confirmation.

She was diagnosed with pre-eclampsia at 20 weeks and was started on oral labetalol 100 mg twice daily.

In the present visit she required in-patient care due to high BP readings of 170/120 mm Hg. Intravenous labetalol injections of 20 mg followed by 40 mg, 80 mg, 80 mg every thirty minutes were administered. Persistent high blood pressure recordings were noted despite giving a maximum of 220 mg of injectable labetalol. Loading dose of injection magnesium sulphate as per Zuspan regimen initiated for neuroprotection with vigilant monitoring. Steroids switched to injection dexamethasone for fetal lung maturity.

Fundoscopy was normal. Endocrinologist opinion sought. Serum cortisol levels sent at 8 AM and 8 PM and plan to restart injection hydrocortisone 50 mg thrice daily after 48 hours of injection dexamethasone was made. Serum Cortisol and serial sugar monitoring was normal.

Despite the maximum dose of injection labetalol, BP was found to be high. Hence, started on labetalol infusion 1.2 mg/min for two hours followed by 60 mg/hour. Screening 2D ECHO done was normal.

Due to persistent high BP recordings despite labetalol infusion, patient was taken for emergency preterm LSCS. Preoperatively, stress dose of injection hydrocortisone 100 mg was administered. A single live male baby was extracted by vertex weighing 1.12 kg at 8:02 AM on 01/08/2024. Intra-operatively, poorly formed Lower uterine segment with reduced liquor was noted. No notable intra-operative complications.

Post-operatively, she was shifted to MICU due to high BP recordings and restarted on Labetalol infusion at 2ml/hour and Injection Magnesium sulphate infusion at 1 gram/hour for 24 hours. MRI brain plain was done and PRES syndrome ruled out. injection hydrocortisone 50 mg thrice a day was administered. IV antibiotics administered prophylactically. On POD 3 patient was shifted out of MICU.

BP values started declining and were controlled with oral amlodipine 5 mg twice daily and clinidipine 10 mg twice daily on post-operative day three. Thromboprophylaxis initiated with low molecular weight heparin. Steroids tapered to injection hydrocortisone 25 mg thrice daily then to tablet prednisolone 20-20-10 mg and then to pre-operative dose and was discharged.

Baby was shifted to NICU for preterm care. Evaluation was done for CAH in the neonate and found to be negative.

No lactational or wound healing issues were noted on follow up.

DISCUSSION

In women with CAH, the problem starts early in life. Classical CAH presents at birth as ambiguous genitalia or salt wasting syndrome while the non-classical form may remain asymptomatic and present later in life from childhood to adolescence with varying degrees of virilization or precocious puberty, hirsutism or infertility.

Pre-conceptional counselling should include treatment options for infertility, optimizing steroid dosage and the probability of having an affected child.[4]

Right dose of the right steroid is the key to fertility. Preferred steroids are hydrocortisone, prednisone and prednisolone since they are metabolized by the placenta.[4]

Management includes genital reconstruction, initiation of appropriate steroids to prevent salt wasting syndrome as well as maintenance of menstruation. Since, establishment of hormonal milieu takes a long time, early initiation results in better conception rates.[4] Psychosexual counselling is an important aspect in the management.

Steroids have to be increased in pregnancy which causes fluid retention, excess weight gain, cushingoid features, hypertension, glucose intolerance and hyperglycaemia.[4]

Placenta acts as a metabolic barrier, reducing the foetal exposure to circulating maternal androgens and its precursor. But in female foetus with Classical CAH, prenatal treatment with dexamethasone to prevent genital virilization is essential.[4]

In women who have undergone genital reconstruction surgery, preferred mode of delivery is LSCS. Administration of stress dose of steroid at the onset of labour to until after delivery is recommended.[4]

In the antepartum period, androgen status is monitored via bioavailable testosterone, which has to be maintained in the high normal range for pregnancy specific reference values.[5,6] Reduction in fertility may be postulated due to reduced amplitude and frequency of luteinising hormone.[5] Challenge here is difficulty in suppression of progesterone or 17-OH-progesterone even with steroid replacement.

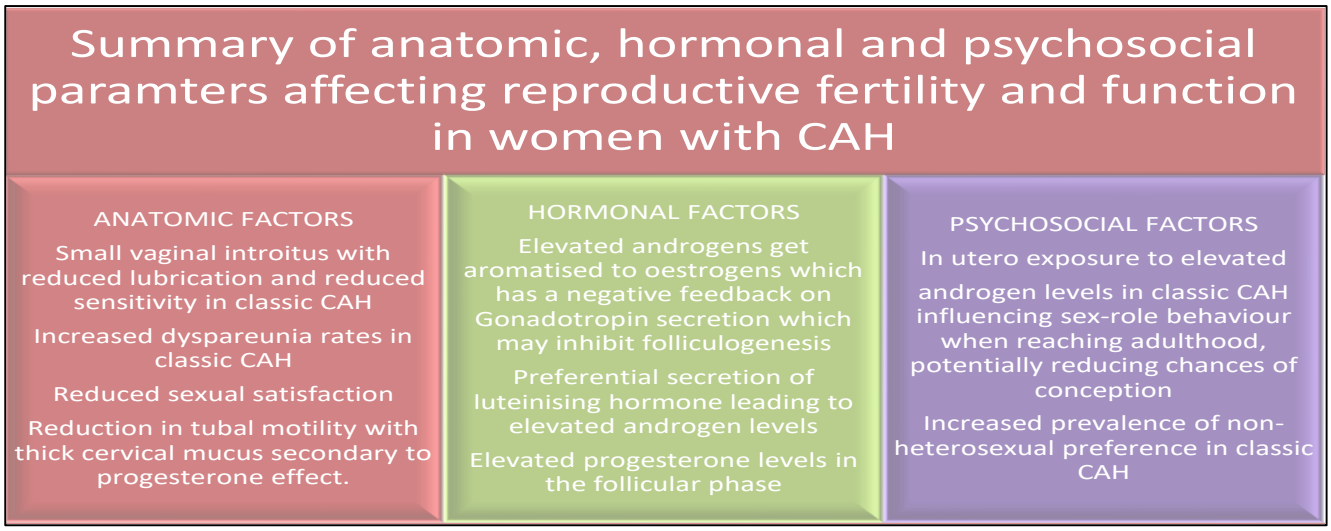

Figure 2: Factors affecting fertility in CAH.[7]

Bilateral adrenalectomy is preferred if excess androgen or progestin is known to cause subfertility but women may continue to have the problem post-surgery, hence it is not preferred.[8]

In few women with infertility assisted reproductive techniques may be suggested.

In our case the CAH was detected early at birth and timely management with steroids and successful reconstructive surgery done. Patient was compliant with the suggested steroid replacement and had regular antenatal visits. She developed pre-eclampsia at 20 weeks and had persistent uncontrolled Hypertension even with Labetalol infusion hence underwent Emergency preterm caesarean delivery. Stress steroid dosing was administered before the caesarean delivery. Post operatively both anti-hypertensives and steroids were tapered.

The neonate was investigated for CAH. Investigations did not reveal CAH in the neonate. Though neonate required NICU care due to preterm delivery, the baby thrived well and was discharged. So far the baby hasn't shown any delay in milestones.

CONCLUSION

In women with classical CAH the emphasis is on early recognition and management. In pregnancy multidisciplinary approach with appropriate steroid replacement is the key. With individualized care, favourable maternal and neonatal outcomes are achievable.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Swamy M, Ganesh D, Ankitha CR. Pregnancy outcome in a woman with congenital adrenal hyperplasia: a rare case report. Int J Reprod Contracept Obstet Gynecol 2026;15:315-8.