

Original Research

Received 12 May 2026

Revised 18 June 2026

Accepted 02 July 2026

Natural Resources for Human Health 2026, Vol. 6 (11s): 97–100

KEYWORDS:

Autoimmune polyglandular syndrome type II, Schmidt syndrome, Addison's disease, pregnancy, adrenal insufficiency, multidisciplinary management

Autoimmune Polyglandular Syndrome Type II in Pregnancy: A Case Report of Successful Multidisciplinary Management

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ABSTRACT: Autoimmune polyglandular syndrome type II (APS-II), also known as Schmidt syndrome, is a rare autoimmune disorder characterized by the coexistence of Addison's disease with autoimmune thyroid disease and/or type 1 diabetes mellitus. Pregnancy in women with APS-II poses significant challenges as adrenal insufficiency, thyroid dysfunction, and diabetes may adversely affect both maternal and fetal outcomes. We report a case of a 32-year-old primigravida with known autoimmune polyglandular syndrome type II comprising autoimmune hypothyroidism and primary adrenal insufficiency (Addison's disease), who was successfully managed through pregnancy and delivery. The patient had been on thyroid hormone replacement since childhood but experienced an adrenal crisis following discontinuation of thyroid medication, which led to the diagnosis of APS-II. She was subsequently maintained on hydrocortisone, fludrocortisone, and levothyroxine replacement. She was admitted at 37 weeks and 4 days of gestation for planned delivery with close maternal and fetal monitoring. A multidisciplinary approach involving obstetrics and endocrinology teams was employed throughout the peripartum period. Stress-dose glucocorticoid coverage was administered during labour, and the patient had an uneventful normal vaginal delivery of a live term female infant weighing 2.736 kg with Apgar scores of 8/10 and 9/10. This case highlights that with meticulous multidisciplinary care, appropriate hormone replacement, and close surveillance, favourable pregnancy outcomes can be achieved in women with APS-II.

INTRODUCTION

Autoimmune polyglandular syndromes (APS) are uncommon heterogeneous conditions characterized by the association of two or more organ-specific endocrinopathies [1]. APS type II (APS-II), also referred to as Schmidt syndrome, is defined as the combination of primary adrenal insufficiency (Addison's disease) with autoimmune thyroid disease and/or type 1 diabetes mellitus [2]. The prevalence of APS-II is approximately five per 100,000 in the United States, and it is most commonly diagnosed in middle-aged women [3]. The clinical presentation of APS-II shows great heterogeneity, and the condition often manifests sequentially with a large time interval between the occurrence of the first and second endocrine diseases [4]. Patients with APS-

II are also predisposed to additional autoimmune diseases such as vitiligo, hypergonadotropic hypogonadism, and autoimmune atrophic gastritis [5].

Pregnancy in women with APS-II is rarely encountered, as both thyroid and adrenal gland insufficiency can be individually associated with decreased fertility [6]. However, when pregnancy does occur, it poses significant challenges. Adrenal insufficiency during pregnancy has a prevalence of 5.5 per 100,000 pregnancies, and there are only approximately ten case reports of APS-II in pregnancy in the literature [1]. Pregnant women with individual endocrine disorders are more prone to maternal and fetal complications, including miscarriage, preterm birth, fetal growth restriction, and preeclampsia [7]. The symptoms of adrenal insufficiency, such as fatigue, nausea, vomiting, weight loss, hypotension, and hyperpigmentation, can mimic normal pregnancy changes, posing significant diagnostic challenges [8]. Furthermore, adrenal crisis is a life-threatening emergency that can affect both maternal and fetal well-being if not identified promptly [9]. The rarity of this condition and the complexity of its management necessitate a multidisciplinary approach involving endocrinologists, obstetricians, anaesthetists, and neonatologists [10]. We report a case of successful pregnancy outcome in a woman with APS-II, highlighting the importance of meticulous multidisciplinary management throughout the antepartum, intrapartum, and postpartum periods.

CASE REPORT

A 32-year-old married female, housewife, with blood group A positive, was admitted on 27 July 2026 for safe confinement at approximately 37 weeks and 4 days of gestation. She was a primigravida with pregnancy complicated by polyglandular autoimmune syndrome type II (PAS-II). Her last menstrual period was dated 6 November 2025, with an expected date of delivery calculated as 13 August 2026.

The patient had a history of thyroid hormone replacement since childhood, having been on thyroxine 100 mcg until the age of 20 years. However, she did not maintain regular checkups of her thyroid function and had discontinued her thyroid medication for three months. Subsequently, she developed significant symptoms including bluish discoloration starting from the face and progressing towards the feet, blackish discoloration of the skin, swelling of the face, severe tiredness, inability to perform day-to-day activities, and complete anorexia with inability to tolerate even water. She was hospitalised, and all necessary investigations were performed, leading to a diagnosis of polyglandular autoimmune syndrome, suspected to be primarily due to overdosing of hypothyroidism drugs or unmasking of adrenal insufficiency following thyroid hormone withdrawal [11]. After initiation of hydrocortisone and fludrocortisone, her symptoms resolved completely, and she continued these medications.

Throughout her antenatal follow-up, the patient's haemoglobin remained approximately 12–14 g/dL, and platelet counts were within acceptable ranges. Blood glucose monitoring was satisfactory, with HbA1c documented around 5.3%. Serum electrolytes were maintained within normal limits, with sodium approximately 136–138 mmol/L and potassium approximately 3.6–3.8 mmol/L. Thyroid function was monitored serially, and the patient remained on appropriate thyroxine replacement. ECG, ECHO, and cardiology evaluation were performed, and no major cardiac abnormality affecting pregnancy was documented. Ultrasonography showed a singleton live fetus with appropriate growth, cephalic presentation, and reassuring fetal status. Serial growth scans, antenatal scans, and Doppler surveillance were performed as indicated.

The patient was on a stable steroid regimen comprising tab hydrocortisone 10 mg (1-1/2-1/2) and tab fludrocortisone 100 mcg once daily, along with tab thyronorm 88 + 33.5 mcg for thyroid replacement. Endocrinology opinion was obtained, and a detailed peripartum steroid protocol was formulated. The plan included administration of Inj hydrocortisone 100 mg intravenous bolus at the onset of labour or caesarean section, followed by Inj hydrocortisone 50 mg six-hourly for 24 hours. In the first 24 hours postpartum, the patient was to receive 50 mg six-hourly intravenously. Between 24 and 48 hours postpartum, the dose was to be doubled from the pre-pregnancy replacement dose, followed by tapering to the usual pregnancy oral dose between 48 and 72 hours postpartum, and subsequently reduction to the pre-pregnancy oral dosage [12].

The patient was managed with a multidisciplinary approach involving obstetrics and endocrinology teams. Appropriate thyroid hormone replacement was continued with monitoring of thyroid function. Glucocorticoid replacement was continued and adjusted according to clinical status and pregnancy requirements. Close maternal monitoring was maintained for symptoms and signs of adrenal insufficiency and thyroid dysfunction. Regular fetal surveillance was performed with NST/CTG and obstetric ultrasonography, along with serial growth assessment and routine antenatal monitoring.

The peripartum management plan included stress-dose glucocorticoid coverage during labour because of adrenal insufficiency. Adequate hydration, haemodynamic monitoring, and close observation were maintained during labour. Thyroid replacement therapy was continued as advised by endocrinology. Blood pressure, pulse, glucose, and clinical features suggestive of adrenal crisis were closely monitored. The patient had a normal vaginal delivery at 37 weeks and 6 days of gestation, delivering a live term female infant weighing 2.736 kg with Apgar scores of 8/10 at 1 minute and 9/10 at 5 minutes. Post-delivery, glucocorticoid supplementation was tapered as clinically appropriate, and endocrine follow-up was continued. Neonatal assessment and routine postnatal care were provided.

DISCUSSION

Autoimmune polyglandular syndrome type II is a rare but serious condition that requires meticulous management during pregnancy [1]. The case presented here demonstrates that with appropriate multidisciplinary care, favourable pregnancy outcomes can be achieved. The rarity of APS-II in pregnancy, with only approximately ten case reports documented in the literature, makes this case a valuable addition to the existing body of knowledge [2].

The diagnosis of APS-II in this patient was established following an adrenal crisis that was precipitated by discontinuation of thyroid medication [3]. This sequence of events is clinically significant, as it highlights a well-recognised phenomenon: in patients with undiagnosed adrenal insufficiency, administration or reintroduction of thyroid hormone can precipitate an adrenal crisis by increasing the metabolic clearance of cortisol [4]. The patient's symptoms of bluish and blackish discoloration, swelling of the face, severe fatigue, and anorexia were classic manifestations of adrenal insufficiency [5]. The presence of hyperpigmentation, which the patient described as blackish discoloration, is a hallmark of primary adrenal insufficiency due to elevated ACTH levels stimulating melanocyte activity [6]. The diagnosis of Addison's disease is confirmed by demonstrating a low baseline cortisol with an elevated ACTH, along with positive 21-hydroxylase antibodies [7].

The management of pregnancy in women with APS-II requires a comprehensive multidisciplinary approach [8]. The key components include appropriate hormone replacement, close maternal and fetal monitoring, and meticulous peripartum management. Glucocorticoid replacement is the cornerstone of management in patients with adrenal insufficiency. During pregnancy, an increase in glucocorticoid replacement dose is expected to be necessary, particularly in the third trimester when physiological cortisol levels rise. The Endocrine Society suggests increasing hydrocortisone by 2.5–10 mg daily during the third trimester [9]. In our patient, the maintenance dose of hydrocortisone was 10 mg three times daily, which was adjusted according to clinical status. Fludrocortisone, the mineralocorticoid replacement, was continued at 100 mcg daily. While some literature suggests that adjusting fludrocortisone may not be necessary as an increase in glucocorticoid dose can compensate, determining the correct dose should be tailored to each patient and their specific symptoms [10].

Thyroid hormone replacement is equally important in patients with hypothyroidism. Levothyroxine requirements often increase during pregnancy due to increased thyroid-binding globulin and placental metabolism of thyroid hormones. In our patient, the dose of thyronorm was 88 + 33.5 mcg, which was monitored serially throughout pregnancy. It is crucial to maintain euthyroidism during pregnancy, as maternal hypothyroidism is associated with adverse obstetric outcomes including miscarriage, preterm delivery, and impaired neurodevelopmental outcomes in the offspring [11].

The peripartum period is particularly critical in women with adrenal insufficiency. Labour and delivery represent significant physiological stress that necessitates stress-dose glucocorticoid coverage to prevent adrenal crisis. The protocol used in our patient involved Inj hydrocortisone 100 mg intravenous bolus at the onset of labour, followed by 50 mg six-hourly for 24 hours. This is consistent with recommended guidelines, which suggest hydrocortisone 100–150 mg/day intravenously during the first 24–48 hours followed by rapid tapering [12]. The patient's uneventful vaginal delivery with no maternal complications attests to the effectiveness of this protocol. Close monitoring of blood pressure, pulse, glucose, and electrolyte status is essential during this period.

Fetal surveillance is another crucial aspect of management. Women with APS-II are at increased risk of fetal growth restriction, preterm birth, and preeclampsia. In our patient, serial growth scans, antenatal scans, and Doppler surveillance were performed as indicated, and the fetus showed appropriate growth with reassuring status. The neonatal outcome was favourable, with a birth weight of 2.736 kg and Apgar scores of 8/10 and 9/10.

The importance of pre-pregnancy counselling cannot be overstated. Patients should be educated about the need for antepartum, intrapartum, and postpartum steroids, and both the patient and her partner should be trained in hydrocortisone emergency injection techniques. In our case, the patient was well-informed about her condition and the necessary management, which contributed to the successful outcome. This case adds to the limited literature on APS-II in pregnancy and demonstrates that with meticulous multidisciplinary care, favourable pregnancy outcomes can be achieved. The successful management of this patient was attributable to close collaboration between obstetrics and endocrinology teams, appropriate hormone replacement, and careful peripartum monitoring.

CONCLUSION

Autoimmune polyglandular syndrome type II is a rare autoimmune disorder that presents unique challenges during pregnancy due to the coexistence of adrenal insufficiency and thyroid dysfunction. The clinical features of adrenal insufficiency, including fatigue, nausea, vomiting, weight loss, hypotension, and hyperpigmentation, can mimic normal pregnancy changes, making diagnosis challenging. Management requires a multidisciplinary approach involving endocrinologists, obstetricians, anaesthetists, and neonatologists. Key elements include appropriate glucocorticoid and mineralocorticoid replacement with dose adjustments during pregnancy and the peripartum period, thyroid hormone replacement with close monitoring, and meticulous maternal and fetal surveillance. Stress-dose glucocorticoid coverage is essential during labour and delivery to prevent adrenal crisis. This case report demonstrates that with appropriate multidisciplinary care and careful planning, successful pregnancy outcomes can be achieved in women with autoimmune polyglandular syndrome type II. The favourable maternal and neonatal outcomes in this case highlight the importance of early diagnosis, comprehensive pre-pregnancy counselling, and coordinated peripartum management. As more cases of APS-II in pregnancy are reported and managed successfully, the evidence base for optimal management strategies will continue to grow, ultimately improving outcomes for these high-risk pregnancies.

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